26th international symposium on ALS/MND

Orlando USA
11 – 13 December 2015

Programme

Host: The ALS Association

Organised by the Motor Neurone Disease Association in co-operation with the International Alliance of ALS/MND Associations
The 26th 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:

The ALS Association
1275 K Street NW, Suite 250
Washington, DC 20005, USA
Tel: (-) 202 407 8580
Fax: (-) 202 289 6801
Email: alsinfo@als-national.org
Website: www.alsa.org

Organiser of the symposium:

Motor Neurone Disease Association
PO Box 246, Northampton NN1 2PR, 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
Fax: (-) 1 215 543 3366
Email: alliance@als-mnd.org
Website: www.alsmndalliance.org

CME Accreditation
The 26th 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

Welcome to Orlando!

The ALS Association is proud to welcome you to the 26th International Symposium on ALS/MND and the accompanying 23rd Annual Meeting of the International Alliance of ALS/MND Associations. Participation in these meetings underscores the global commitment of each and every attendee to make a difference in the fight against ALS.

Orlando, nicknamed ‘The City Beautiful’, is a wonderful location to convene and share the latest developments in scientific research, along with care and support for people living with ALS.

The ALS Association, which includes 39 chapters working in all fifty states across the United States, has an integrated mission that includes research, care services and public policy, which all aim to find treatments and a cure for ALS.

There has never been a more exciting time in ALS research. The ALS Association is currently funding 125 active projects in 12 countries, including four new global collaborative initiatives to build understanding of the disease, target new therapies, expedite clinical trials, and make DNA and RNA sequencing data available to the entire ALS research community.

Within the past year, scientists funded by The ALS Association have made important discoveries around ALS gene mutations, leading to three new academic-industry contracts in our drug development program.

There is also significant progress in the development of antisense technology for neurodegenerative diseases such as ALS. This progress can be directly attributed to The ALS Association’s and MDA’s early and continued investments in this area. We expect antisense clinical trials to occur in 2016 and early 2017.

Foreword

Welcome to the 26th International Symposium on ALS/MND. The next three days will offer a chance to hear about the latest scientific discoveries and advances in the delivery of high quality care for people living with ALS. The breathtaking pace at which laboratory science is revealing the molecular and cellular events behind motor neuron degeneration is exciting, but also serves to remind us that we are dealing with a disease of immense biological complexity. We now have a large number of different genes and pathways to study, but translating these into effective therapies to modify disease progression and to enhance wellbeing remains challenging. The scientific presentations at this meeting, by taking us from the basic mechanisms of disease at the RNA and protein level through to the latest advances in targeted biological therapy will provide a road map for the future treatment of ALS.

Over the years, The ALS Association has been pleased to partner with industry, government and with other ALS organizations. With increased research revenue, we have had the opportunity to expand these partnerships globally, accelerating drug development so that people living with the disease will be closer to receiving meaningful treatments.

People living with ALS are at the center of everything we do. In the past year, The ALS Association has expanded access to care in underserved communities while also doubling our grants to Certified Treatment Centers of Excellence so that patients can receive the very best multidisciplinary care and participate in cutting-edge research.

Learning from our colleagues in Europe, The ALS Association is in the midst of an ambitious initiative to expedite drug development, working with the U.S. Food and Drug Administration to create a guidance document to help companies navigate the regulatory pathway.

We look forward to talking more about these and other important initiatives during our time in Orlando, which is also known as the ‘Theme Park Capital of the World’. While you’re here, you may find yourself humming the popular song, ‘It’s a small world after all’, an appropriate anthem for the collaborative spirit of these meetings.

Together, we will end this disease.

Barbara J. Newhouse
President and CEO,
The ALS Association

Advances in care and symptom management also have their foundation in carefully conducted evidenced-based research. The benefits of high quality symptom management, nutritional and respiratory support through a multidisciplinary team effort seem self-evident. But when resources and expertise are limited we owe it to those living with ALS/MND to choose our interventions wisely and use the best evidence available. As we will learn at this meeting, good research and good care go hand in hand.

On behalf of the Programme Committee I wish you a happy and productive meeting.

Prof Kevin Talbot
Programme Committee Chair
## Programme

### Friday 11 December 2015

#### SESSION 1  JOINT OPENING SESSION

**Chairs:**

- S Light (UK)
- K Talbot (UK)

<table>
<thead>
<tr>
<th>Time</th>
<th>Event</th>
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<tbody>
<tr>
<td>08.30 – 08.35</td>
<td>Welcome – S Light (UK), K Talbot (UK)</td>
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<tr>
<td>08.35 – 08.40</td>
<td>Welcome – Barbara Newhouse, President and CEO, The ALS Association (USA)</td>
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<tr>
<td>08.40 – 09.10</td>
<td>Addressing the burden of neurodegenerative disease: a global challenge – R Shakir (UK)</td>
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#### SESSION 2A  RNA BIOGENESIS AND PROCESSING

**Chairs:**

- J Robertson (Canada)
- S McKnight (USA)

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<tr>
<td>11.00 – 11.30</td>
<td>The role of low complexity domains in proteins involved in ALS – S McKnight (USA)</td>
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<tr>
<td>11.30 – 11.50</td>
<td>Gain of toxicity from hexanucleotide expansion in C9orf72 in ALS and frontotemporal dementia is alleviated by antisense oligonucleotides targeting repeat-containing RNAs – C Lagier-Tourenne (USA)</td>
</tr>
<tr>
<td>11.50 – 12.10</td>
<td>Neuropathology in C9orf72-ALS is consistent with PURA sequestration and loss of function in motor neurons despite compensatory overexpression – J Cooper-Knock (Sheffield)</td>
</tr>
<tr>
<td>12.10 – 12.30</td>
<td>RBM4S homo-oligomerization mediates association with ALS-linked proteins and stress granules – Y Li (USA)</td>
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#### SESSION 2B  CLINICAL MANAGEMENT

**Chairs:**

- O Hardiman (Ireland)
- V Silani (Italy)

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<td>11.00 – 11.30</td>
<td>Multidisciplinary care in ALS: measuring the immeasurable? – O Hardiman (Ireland)</td>
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<td>The experimental treatment of bulbar symptoms in ALS with Nuedexta – R Smith (USA)</td>
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<td>Treatment of medically refractory sialorrhea with electron beam radiotherapy (EBRT) to the parotid – E Kasarskis (USA)</td>
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<td>A national study of muscle cramps in amyotrophic lateral sclerosis – B Oskarsson (USA)</td>
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#### SESSION 3A  RNA LOCALIZATION AND DYSREGULATION

**Chairs:**

- J Rothstein (USA)
- J Taylor (USA)

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<tbody>
<tr>
<td>14.00 – 14.30</td>
<td>Altered RNA Metabolism in ALS and related diseases – J Taylor (USA)</td>
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<tr>
<td>14.30 – 14.50</td>
<td>Nuclear pore abnormalities in C9orf72 ALS iPS neurons and tissue alter nucleocytoplasmic protein trafficking – C Donnelly (USA)</td>
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<tr>
<td>14.50 – 15.10</td>
<td>Characterization of C9orf72 expression in pathological cohort uncovers new clinical associations with specific C9orf72 transcripts – M Van Blitterswijk (USA)</td>
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<tr>
<td>15.10 – 15.30</td>
<td>In-depth analysis of sense and antisense RNA foci in a large pathological cohort of C9orf72 expansion carriers – M DeJesus-Hernandez (USA)</td>
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#### SESSION 3B  HOLISTIC CARE

**Chairs:**

- K Pearce (UK)
- J Armstrong (USA)

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<td>14.00 – 14.30</td>
<td>Discussing prognosis with ALS/MND patients: Balancing hope and realism – J Jacobsen (USA)</td>
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<td>14.30 – 14.50</td>
<td>Experiences of people with Motor Neurone Disease (MND) and their family caregivers receiving the news of diagnosis: A national survey – S Aoun (Australia)</td>
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<td>14.50 – 15.10</td>
<td>The impact of family on decision-making in ALS care: The patient perspective – G Foley (Ireland)</td>
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<td>15.10 – 15.30</td>
<td>Determinants of caregiver strain in carers of patients with ALS: A longitudinal study – H Creemers (The Netherlands)</td>
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**Reception at Sunset**

Location: Mediterranean 5

**Monday 14 December 2015**

#### SESSION 1A  RNA BIOGENESIS AND PROCESSING

**Chairs:**

- M Behrens (Canada)
- S McKnight (USA)

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#### SESSION 3B  CLINICAL MANAGEMENT

**Chairs:**

- O Hardiman (Ireland)
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**Reception at Sunset**

Location: Mediterranean 5
### SESSION 4A: DISEASE MODELS

**Chairs:** S Hadano (Japan) L Van Den Bosch (Belgium)

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<thead>
<tr>
<th>Time</th>
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<th>Speaker/Country</th>
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<tbody>
<tr>
<td>16.00 – 16.30</td>
<td>Generating new mouse models of ALS – C Lutz (USA)</td>
<td>C Lutz (USA)</td>
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<tr>
<td>16.30 – 16.50</td>
<td>New mouse models of ALS show neuron survival and motor recovery after clearance of TDP-43 pathology – A Walker (USA)</td>
<td>A Walker (USA)</td>
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<tr>
<td>16.50 – 17.10</td>
<td>A gain of toxicity by C9orf72 GGGGCC expansion in amyotrophic lateral sclerosis and frontotemporal dementia – Q Zhu (USA)</td>
<td>Q Zhu (USA)</td>
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<tr>
<td>17.10 – 17.30</td>
<td>An in vivo model of BMAA-induced protein inclusions of Guam ALS/PDC – P Cox (USA)</td>
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### POSTER SESSION A

**Location:** Coquina North

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<thead>
<tr>
<th>Time</th>
<th>Theme</th>
<th>Duration</th>
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<tbody>
<tr>
<td>18.00 – 18.20</td>
<td>Clinical work in progress and care practice</td>
<td>20 minutes</td>
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<tr>
<td>18.20 – 18.40</td>
<td>Molecular cell biology</td>
<td>20 minutes</td>
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<tr>
<td>18.40 – 19.00</td>
<td>ALS heterogeneity and disease progression</td>
<td>20 minutes</td>
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<tr>
<td>19.00 – 19.20</td>
<td>Clinical management</td>
<td>20 minutes</td>
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<tr>
<td>19.20 – 19.40</td>
<td>In vitro experimental models</td>
<td>20 minutes</td>
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### Saturday 12 December 2015

**SESSION 5A: GENE THERAPY**

**Chairs:** C Svendsen (USA) N Maragakis (USA)

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<tbody>
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<td>08.30 – 09.00</td>
<td>Targeting RNA mis-splicing in SMA – A Krainer (USA)</td>
<td>A Krainer (USA)</td>
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<tr>
<td>09.00 – 09.30</td>
<td>Developing therapeutics for neuromuscular disease: from basic to translational studies – B Kaspar (USA)</td>
<td>B Kaspar (USA)</td>
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<tr>
<td>09.30 – 09.50</td>
<td>A new AAV-based gene therapy approach for SOD1-linked ALS – M G Biferi (France)</td>
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**SESSION 5B: GENETIC COUNSELLING**

**Chairs:** P Andersen (Sweden) A Chiò (Italy)

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<td>08.30 – 09.00</td>
<td>ALS Genetics: Helping your patients and families understand – N Siddique (USA)</td>
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<tr>
<td>09.00 – 09.30</td>
<td>Genetic counselling in the post genomic world – a clinician’s perspective – C Shaw (UK)</td>
<td>C Shaw (UK)</td>
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<tr>
<td>09.30 – 09.50</td>
<td>Pre-symptomatic ALS genetic counselling and testing: Experience and recommendations – M Benatar (USA)</td>
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**10.00 – 10.30 COFFEE** Location: Mediterranean Foyer
**SESSION 6A**

**CLINICAL-PATHOLOGICAL CORRELATES OF DISEASE PROGRESSION**

**Chairs:**
- J Ravits (USA)
- M Kiernan (Australia)

*Location: Mediterranean 6-8*

**10.30 – 11.00**
Understanding disease progression in ALS – J Ravits (USA)

**11.00 – 11.30**
Prions in neurodegenerative diseases: amyloid structures dictate disease characteristics – M Diamond (USA)

**11.30 – 12.00**
Frontotemporal dementia: onset, spread and relationship to ALS – B Seeley (USA)

**12.00 – 12.20**
TDP-43 proteinopathies: pathological identification of brain regions differentiating clinical phenotypes – R Tan (Australia)

**12.20 – 12.40**
Correlation of pathological and clinical phenotypes in C9orf72 carriers – J Scaber (UK)

**12.30 – 14.00 LUNCH**

**SESSION 6B**

**RESPIRATORY SUPPORT**

**Chairs:**
- J Rosenfeld (USA)
- PN Leigh (UK)

*Location: Mediterranean 5*

**10.30 – 11.00**
Living and dying with invasive home mechanical ventilation in patients with advanced ALS: decision-making, survival and withdrawal – P Dreyer (Denmark)

**11.00 – 11.30**
Permanent ventilation via tracheostomy: a one-way street? – M Davies (UK)

**11.30 – 11.50**
The development of guidance for professionals in the UK for the withdrawal of assisted ventilation at the request of a patient with MND – C McDermott (UK)

**11.50 – 12.10**
Multi-center diaphragm pacing post FDA approval study enrolment complete: Favorable procedural success, interim safety and survival findings – R Miller (USA)

**12.10 – 12.30**
A randomised controlled trial of the effectiveness of the NeuRx RA/4 diaphragm pacing system in patients with respiratory muscle weakness due to amyotrophic lateral sclerosis (ALS) (The DiPALS Trial) – C McDermott (UK)

**SESSION 7A**

**GENETICS AND EPIGENETICS**

**Chairs:**
- F Baas (The Netherlands)
- W Camu (France)

*Location: Mediterranean 6-8*

**14.00 – 14.30**
Distinct brain transcriptomes and methylomes in C9orf72-associated and sporadic ALS – L Petrucelli (USA)

**14.30 – 14.50**
Genetic overlap between amyotrophic lateral sclerosis and schizophrenia – R McLaughlin (Ireland)

**14.50 – 15.10**
Large scale genetic screening in sporadic ALS identifies modifiers in C9orf72 repeat carriers – M Van Es (The Netherlands)

**15.10 – 15.30**
Exome sequencing of familial ALS index cases identifies a novel ALS gene that binds calcium – B Smith (UK)

**SESSION 7B**

**NON-MOTOR SYMPTOMS OF ALS**

**Chairs:**
- R Sufit (USA)
- G Mora (Italy)

*Location: Mediterranean 5*

**14.00 – 14.30**
Pseudobulbar affect: From biology to management – J Parvizi (USA)

**14.30 – 14.50**
I can’t help that I look sad: the experience of emotional liability in the ALS patient and caregiver – Z Adirim (Australia)

**14.50 – 15.10**
Apathy and executive dysfunction in ALS – R Radakovic (UK)

**15.10 – 15.30**
Differentiating depression from PBA-related crying in ALS: Use of PHQ-9 and CNS-LS – N Thakore (USA)

**SESSION 7C**

**BIOMARKERS**

**Chairs:**
- E Pioro (USA)
- N Atassi (USA)

*Location: Mediterranean 1-2*

**14.00 – 14.30**
Advances in PET imaging in ALS and related conditions – N Atassi (USA)

**14.30 – 14.50**
Metabolic spatial connectivity in Amyotrophic lateral sclerosis: A 18-FDG PET study – A Chiò (Italy)

**14.50 – 15.10**
Retinal thinning in amyotrophic lateral sclerosis: A study with optical coherence tomography and diffusion tensor imaging – A Hübers (Germany)

**15.10 – 15.30**
The potential of whole-body muscle MR as a biomarker in amyotrophic lateral sclerosis – T Jenkins (UK)

**15.30 – 16.00 COFFEE**

*Location: Mediterranean Foyer*
MECHANISMS OF INTERCELLULAR PROPAGATION

Chairs: T Brännström (Sweden) N Cashman (Canada)

16.00 – 16.20 Translational profiling identifies a cascade of damage that initiates in motor neurons and spreads to glia in mutant SOD1-mediated ALS – S Sun (USA)

16.20 – 16.40 Propagation of SOD1 misfolding in an ALS mouse model – J Ayers (USA)

16.40 – 17.00 Use of a fluorescently-tagged permissive substrate reporter system to detect SOD1 prion-like activity in living cells: Implications for an ALS therapeutic drug screen – E Pokrishevsky (Canada)

17.00 – 17.20 TDP-43 prionoids trigger ALS-associated pathology in normal mouse brain slices – M Polymenidou (Switzerland)

17.20 – 17.40 TDP-43 is released in association with exosomes – Y Iguchi (Canada)

NUTRITIONAL AND RESPIRATORY ASSESSMENT AND INTERVENTION

Chairs: B Brooks (USA) E Kasarkskis (USA)

16.00 – 16.20 Weight loss at diagnosis and survival in ALS: A population-based study – B Marin (France)

16.20 – 16.40 Hydration status, disease progression and survival in ALS patients – R Tandan (USA)

16.40 – 17.00 A risk-stratifying tool to facilitate safe late-stage percutaneous endoscopic gastrostomy in MND – A Thompson (UK)

17.00 – 17.20 Discriminate ability of the eating assessment tool for predicting aspirations – E Plowman (USA)

17.20 – 17.40 A randomised sham control trial of EMST on bulbar function – E Plowman (USA)

ELECTROPHYSIOLOGY AND IMAGING

Chairs: C Neuwirth (Switzerland) M Ross (USA)

16.00 – 16.20 MUNIX detects presymptomatic motor unit loss in ALS patients – C Neuwirth (Switzerland)

16.20 – 16.40 The natural history of motor cortical function in amyotrophic lateral sclerosis – K Shibuya (Australia)

16.40 – 17.00 Riluzole exerts short-term effects on cortical hyperexcitability in sporadic ALS – N Geevasinga (Australia)

17.00 – 17.20 Effects of the C9orf72 repeat expansion: a neuroimaging investigation of brain morphological changes in asymptomatic mutation carriers – R Walhout (The Netherlands)

17.20 – 17.40 Structural and functional MRI signatures of ALS patients with C9orf72 hexanucleotide repeat expansion – F Agosta (Italy)

BIOMARKERS AND OUTCOME MEASURES

Theme 3: Biomarkers and outcome measures

Epidemiology

Imaging and electrophysiology

ALS pathogenesis and neurotoxicity

Cognitive change

Therapeutic strategies

Quality of life and palliative care

Scientific work in progress
## Sunday 13 December 2015

### SESSION 9A  
**MOTOR NEURON VULNERABILITY**

**Location:** Mediterranean 6-8  
**Chairs:**  
- M Sendtner (Germany)  
- S Burden (USA)

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<td>08.30 – 09.00</td>
<td>Dysregulation of axonal RNA processing in motor neuron disease – M Sendtner (Germany)</td>
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<tr>
<td>09.00 – 09.30</td>
<td>Preserving neuromuscular synapses in ALS – S Burden (USA)</td>
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<tr>
<td>09.30 – 10.00</td>
<td>Connecting genes to pathways and networks in an effort to reveal the basis of selective motor neuron vulnerability – H Ozdinler (USA)</td>
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### SESSION 9B  
**CLINICAL TRIALS**

**Location:** Mediterranean 5  
**Chairs:**  
- A Ludolph (Germany)  
- J Shefner (USA)

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<td>08.30 – 09.00</td>
<td>Confirmatory double-blind, parallel-group, placebo-controlled study of efficacy and safety of edaravone (MCI-186) in ALS patients – Edaravone (MCI-186) ALS Study Group (Japan)</td>
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<tr>
<td>09.00 – 09.20</td>
<td>Adaptive design single center phosphodiesterase type 4 (PDE4) inhibitor: Phase 1b/2a clinical trial – B Brooks (USA)</td>
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<td>09.20 – 09.40</td>
<td>Ultra-high dose methylcobalamin (E0302) prolongs survival of ALS by more than 600 days if treated early: Randomized double-blind, phase 3 clinical trial (ClinicalTrials.gov NCT00444613) – R Kaji (Japan)</td>
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<tr>
<td>09.40 – 10.00</td>
<td>A phase 2 study for safety and efficacy evaluation of treatment of amyotrophic lateral sclerosis using autologous bone-marrow-derived stromal cell – S H Kim (Republic of Korea)</td>
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### 10.00 – 10.30 COFFEE  
**Location:** Mediterranean Foyer

### SESSION 10A  
**DISEASE MECHANISMS AND THERAPEUTIC TARGETS**

**Location:** Mediterranean 6-8  
**Chairs:**  
- B Turner (Australia)  
- J Prehn (Ireland)

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<td>Direct conversion as a method to subgroup ALS patient populations – K Meyer (USA)</td>
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<td>10.50 – 11.10</td>
<td>A transcriptional comparison of human iPSC and mouse models of ALS defines the impact of motor neuron maturation, aging and disease – R Ho (USA)</td>
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<td>Using C. Elegans to identify conserved modifiers of C9orf72-associated dipeptide toxicity – T Lamitina (USA)</td>
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<td>In vivo chemigenetics reveals neuroprotective excitation-related signalling in neurons and astrocytes in SOD1(G93A) mouse – F Roselli (Germany)</td>
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<td>Bromodomain inhibitors regulate the C9orf72 locus in ALS – Z Zeier (USA)</td>
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<td>Genetic removal of histone deacetylase 6 (Hdac6) delays the disease progression in a FUS mouse model of ALS – E Pollari (Belgium)</td>
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<td>12.30 – 12.50</td>
<td>Protection by copper delivery in SOD-transgenic mice and the importance of the Copper Chaperone for SOD1 (CCS) – J Beckman (USA)</td>
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### SESSION 10B  
**COGNITIVE CHANGE**

**Location:** Mediterranean 5  
**Chairs:**  
- M Strong (Canada)  
- M Hornberger (UK)

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<tr>
<td>11.00 – 11.40</td>
<td>The social brain of patients with amyotrophic lateral sclerosis (ALS): the more the better – H Aho-Oezhan (Germany)</td>
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<td>11.20 – 12.00</td>
<td>Relationship between cognitive and behavioural impairment and depression in a large ALS cohort – J Rabkin (USA)</td>
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<td>12.00 – 12.20</td>
<td>Neuropsychiatric symptoms predictive of greater caregiver distress in ALS – M Hornberger (UK)</td>
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### 12.30 – 14.00 LUNCH  
**Location:** Coquina South

### SESSION 11  
**JOINT CLOSING SESSION**

**Location:** Mediterranean 5-8  
**Chairs:**  
- K Talbot (UK)  
- L Bruijn (USA)

<table>
<thead>
<tr>
<th>Time</th>
<th>Session</th>
<th>Speaker(s)</th>
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<tbody>
<tr>
<td>14.00 – 14.05</td>
<td>Poster Prize presentation</td>
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<tr>
<td>14.05 – 14.10</td>
<td>Invitation to Dublin 2016</td>
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<td>14.10 – 14.40</td>
<td>Stem cell treatment strategies for neurodegenerative disease: from hype to hope? – C Svendsen (USA)</td>
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<td>14.40 – 15.10</td>
<td>Patient 3.0: the centre of drug development – B Muller (The Netherlands)</td>
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<tr>
<td>15.10 – 15.20</td>
<td>Late breaking news</td>
<td></td>
</tr>
</tbody>
</table>
Poster sessions

THEME 1
Clinical Management

P1 Clinical diagnosis and multidisciplinary management in amyotrophic lateral sclerosis (ALS)
Santos Salvioni CC, Stanich P, Oda Al, Vital de Carvalho E, Alves PCL, Silva R, Borges RM, Oliveira ASB

P2 Evaluation of the application of the European guidelines for diagnosis and clinical care of ALS patients

P3 Revisiting early diagnosis in ALS
Geevasinga N, Menon P, Loy C, Yiannakis C, Simon N, Kiernan M, Vucic S

P4 Breaking the news of an MND diagnosis: A survey of neurologists in Australia

P5 Familial amyotrophic lateral sclerosis – what motivates first-degree relatives to participate in a preclinical study and how is the study process perceived?

P6 The spectrum of clinical opinion on genetic testing in ALS

P7 The structure and use of the Amyotrophic Lateral Sclerosis Functional Rating Scale revised
Young C, Tennant A on behalf of the Tonic Group

P8 A proposed curriculum for Motor Neurone Disease Specialist Nurses
Burns A, Tuttle L, Lang C, Jung A

P9 Adherence to the Clinical Practice Guideline for Physiotherapy to Patients with ALS in Denmark: A cross-sectional study
Verstere S, Buus L

P10 Coin rotation task for assessing manual dexterity in ALS

P11 Systematic investigation on needs and use of technological devices in Amyotrophic Lateral Sclerosis patients and caregivers: a nationwide, multicentre survey

P12 Assistive equipment use by people with ALS/MND in Australia
Conners K, Mahony L, Mathers S, Morgan P, McPhee M, Solomon S, Smith A

P13 Electronically Augmented Timed Up and Go Test (EATUG) to evaluate mobility and balance in ambulatory patients with amyotrophic lateral sclerosis (ambALS)

P14 Telephone interviews on respite and communication resources to supporting hospitals and people with ALS in a prefecture of Japan
Narita Y, Nakai M

P15 Did morphine usage become more popular in Japan? Based on the findings from 2015 nationwide survey
Ogino M, Tominaga N, Uchino A, Takahashi K, Nagashima K, Yanagita K, Ogino Y

THEME 2
ALS Heterogeneity and Disease Progression

P16 Distinctive pathological features of the Cu/Zn superoxide dismutase 1 (SOD1) mutation D102N
Pallebage-Gamarallage M, Morrison K, Talbot K, Ansorge O

P17 Globular glial tauopathy (type II) clinically mimicking ALS
Tanaka H, Toyoshima Y, Takahashi H

P18 Globular glial mixed four repeat Tau and TDP-43 proteinopathy with motor neuron disease and frontotemporal dementia

P19 Tau oligomer accumulation in patients with globular glial tauopathy (GTT) type III

P20 Elucidating the potential role of antecedent disease in amyotrophic lateral sclerosis

P21 SOD1 mutation mimics a distal hereditary motor neuropathy

P22 Concurrent motor neuron disease and sporadic Parkinson’s disease: two clinical cases of Bright-Fahn-Schwartz complex. Functional assessment and cytokine assays
Dolciotti C, Cavalli L, Ravaiolli S, Paolocci A, Rossi B, Bongianni P

P23 The ALS Stratification Prize – using the power of big data and crowdsourcing for catalyzing breakthroughs in amyotrophic lateral sclerosis

P24 Characteristics associated with decline of anthropometric measurements in ALS patients
Almeida C, Stanich P, Bassoli L, Salvioni C, Oda Al, Dicci C

P25 Correlation between progression rate, need for external aids and survival in patients with ALS
Diaz-Gomez MF, Ortiz-Corredor F, Peña-Preciado M

P26 Medication exposure and survival in ALS: An observational study using propensity score matching
Thakore NJ, Fan Y, Pioro EP

P27 Multi-step analysis shows that late onset ALS with slow progression may represent a distinct aetiological subgroup

P28 Clinical characteristics of elderly patients with amyotrophic lateral sclerosis
Doriey VE, Arntzen J, Vainer B, Nefussy B

P29 Hematological prognostic markers of survival in Dutch amyotrophic lateral sclerosis patients
Visser AE, Westeneng HJ, Veldink JH, van den Berg LH

P30 Creatine kinase enzyme level correlates positively with the serum creatinine and lean body mass, and is a prognostic factor for survival in ALS
Rafaj M, Lee E, Bradburn M, McDermott C, Shaw PJ

P31 Vitamin D: not protective in ALS

P32 Pitfalls in the use of ALSFRS slope in therapeutic trials, and its variable relationship to symptom onset
Proudfoot M, Jones A, Talbot K, Al-Chalabi A, Turner P

P33 VALUES: The effects of low estrogen on executive functioning in a national sample of ALS individuals suggests a neuroendocrine model of disease onset

P34 Staging communication ability in advanced ALS patients

P35 Clinical assessment protocol of the orofacial musculature, with gravity and fatigue markers, targeted to patients with motor neuron disease
Oda Al, Silva R, Alves PCL, Oliveira ASB

THEME 3
Biomarkers and Outcome Measures

P36 Panel of oxidative stress and inflammation markers in sporadic amyotrophic lateral sclerosis

P37 Assessment of a multiple biomarker panel and its combined use for amyotrophic lateral sclerosis
Shang H, Chen X, Chen Y, Wei Q

P38 Decreased level of serum autoantibody against G72 is a biosignature of amyotrophic lateral sclerosis
Hwang C-S, Tsai C-H, Liu G-T, Chang H-T

P39 Protective search for early biomarkers for amyotrophic lateral sclerosis

P40 POSTER WITHDRAWN

P41 The extracellular domain of p75 neurotrophin receptor is present in both urine and cerebrospinal fluid of people living with amyotrophic lateral sclerosis
Ferruaudo L, Shephered S, Schultz D, Chataway T, Wuu J, Benatar M, Shaw PJ, Rogers M-L
P42 Longitudinal changes in urinary p75 neurotrophin receptor extracellular domain levels as a disease progression biomarker in ALS patients

P43 Increased TDP-43 in the skin of patients with amyotrophic lateral sclerosis
Fujikura M, Ono S

P44 Methodological comparison of extracellular vesicle extraction as a new source of biomarkers in ALS

P45 Aberration of MicroRNA Expression in leukocytes from sporadic amyotrophic lateral sclerosis
Chen Y, Wei Q-Q, Li C, Chen X, Shang H

P46 miRNA expression differences in exosomes from identical twins with the C9orf72 repeat expansion discordant for ALS
Schneider R, McKeever P, Petrescu N, Rogaea E, Tartaglia C, Zinnan L, Robertson J

P47 Multi-frequency electrical impedance myography of the tongue: a biomarker for bulbar dysfunction?
Mclduff C, Yim S, Pacheck A, Rutkove S

P48 Comparison of normalization methods in the analysis of maximum voluntary isometric contraction (MVIC) in the EMPower trial
Atassi N, Liu D, Leinet M, Ferguson T, Cerarbaum J, Johns D

P49 Modeling EMPower data to improve the efficiency of ALS phase II/III clinical trials
Leinet ML, MacDougal J, Liu D, Ferguson TA, Cederbaum J, Johns DR, Atassi N

P50 Serum creatinine is a reliable marker for disease severity and prognosis in amyotrophic lateral sclerosis: New evidence based on intrinsic brain activity study
Zhu W, Ren Y, Huang X

P51 Impaired muscle uptake of creatine in spinal and bulbar muscular atrophy

P51A A functional scale for spinal and bulbar muscular atrophy: Cross-sectional and longitudinal study
Hashizume A, Katsumo M, Suzuki K, Hijikata Y, Yamada S, Inagaki T, Sobue G

P52 Longitudinal assessment of reachable workspace in ALS, using the Kinect system

P53 TGF-beta parallels Smad expression in ALS muscle and is a marker of disease progression

THEME 4

Imaging and Electrophysiology

P54 Biomarkers save time, explain mode of action, and add safety in long-term individualized G-CSF compassionate use for ALS patients

P55 Can an MRI-biomarker provide stabilization in G-CSF-treated ALS patients?

P56 Quality control of multicenter, high resolution T1 MRI as a biomarker in amyotrophic lateral sclerosis

P57 MRI correlates of (11C) PB28 PET as a biomarker for ALS

P58 Increased functional connectivity within the sensorimotor resting-state network in ALS: a candidate MEG-based biomarker

P59 The Canadian ALS Neuroimaging Consortium (CALSNC)

P60 A connectivity-based analysis of frontotostial and cortico-basal networks in ALS
Bede P, Schuster C, Elamin M, McLaughlin R, Kenna K, Hardiman O

P61 Whole-brain functional connectivity changes in classical ALS are related to physical disability

P62 Neurite Orientation Dispersion and Density Imaging (NODDI) demonstrates microstructural changes associated with amyotrophic lateral sclerosis
Broad R, Gabel M, Cercignani M, Leigh PN

P63 The selective anatomical vulnerability of ALS - “disease-defining” and “disease defying” brain regions
Bede P, Schuster C, Elamin M, McLaughlin R, Kenna K, Hardiman O

P64 Gray and White matter alteration in ALS patients with or without cognitive impairment: A combined tract based statistical analysis and voxel based morphometry study

P65 Neuronal activation of behaviorally impaired patients with amyotrophic lateral sclerosis in tasks of executive functioning: A functional magnetic resonance imaging study

P66 Post mortem brain imaging to interpret the in vivo MRI signature of MND

P67 Correlation between MRI measures of grey and white matter and clinical measures in C9orf72 symptomatic and asymptomatic carriers
Floeter M, Bageac D, Danielian L, Braun L, Traynor B

P68 Basal ganglia pathology is associated with neuropsychological deficits in C9orf72-negative ALS

P69 Sequential PET and MRI scans in a patient with ALS-FTD reveal worsening brain metabolism and cortical thinning with disease progression
Pioro E, Rajagopalan V

P70 In vivo DTI shows pathology spreading in ALS: staging analysis in more than 300 data sets
Kassubek J, Muller H-P, Del Tredici K, Lulé D, Keller J, Braak H, Ludolph AC

P71 Phenotype-specific white matter signatures in ALS
Schuster C, Elamin M, Hardiman O, Bede P

P72 A three time point, longitudinal imaging analysis in ALS

P73 Apparent diffusion coefficients distinguish amyotrophic lateral sclerosis from cervical spondylotic myelopathy

P74 Structural brain MRI abnormalities in Kennedy's disease
Ferraro PM, Agosta F, Querin G, Riva N, Bertolin C, Da Re E, Copetti M, Comi G, Falini A, Soraru G, Fillippi M

P76 Electromyographic findings of progressive muscular atrophy: Comparison with amyotrophic lateral sclerosis
Bokuda K, Shimizu T, Kimura H, Yamazaki T, Kameyama T, Isozaki E

P77 Power analysis of electroencephalographic mu rhythm in patients with ALS
Malečí T, Kontrnik B, Zidari I, Benlot R, Zidar J

P78 Exploring motor imagery and motor cortical function in amyotrophic lateral sclerosis using magnetoencephalography
Lee M, Meng D, Kiernan M, Johnson B

P79 Sub-threshold repeat length in C9orf72 correlates with brain-computer interface performance
Geronimo A, Simmons Z, Schiff S

P80 Using electrical impedance myography to predict force output in ALS: a study in the G93A SOD1 mouse
Li J, Sanchez B, Pacheck A, Rutkove S

P81 The needle electromyography features in 112 patients with amyotrophic lateral sclerosis
Ren Y, Chen Z, Ling L, Huang X

P82 Evaluation of MUNIX measurements in ALS patients as clinical routine procedure in a specialized neuromuscular treatment unit
Ringer TM, Appelfeller M, Gunkel A, Prell T, Stubendorff B, Witte OW, Grosskreutz J

P83 Increasing weakness... does MRI show anything? Lessons from single subject analysis
Gallagher T, Ajroud-Driss S, Siddique T

P84 The split-hand sign in amyotrophic lateral sclerosis: Different F-wave characteristics between the median and ulnar nerves
Fang J, Cui LY, Liu MS, Guan YZ, Li XG

P85 POSTER WITHDRAWN

P86 POSTER WITHDRAWN
P131 Research priorities in palliative and end of life care: MND results from a broad analysis of patient views

P132 Transitional care for 100 patients with amyotrophic lateral sclerosis
Ye CH, Zheng MY, Shi L, He RJ, Dai JY, He BX, Liu Q, Su HX, Yao XL

P133 The role of palliative care in a multi-disciplinary ALS clinic
Kaplan L, Shahbazi M, Holzberg S, Lange D

P134 Death with dignity in Washington and Oregon patients with amyotrophic lateral sclerosis

THEME 8
Epidemiology

P135 Update on the United States National Amyotrophic Lateral Sclerosis (ALS) Registry
Mehta P, Antavo V, Horton K

P136 National Amyotrophic Lateral Sclerosis (ALS) Biorepository Feasibility Study
Kaye W, Wagner L, Mehta P

P137 National Amyotrophic Lateral Sclerosis (ALS) Registry: A model for recruiting patients for research, clinical trials, and epidemiological studies
Mehta P, Antavo V, Horton K

P138 Incidence of ALS in British Columbia, Canada: A 5-year retrospective study
Golby R, Fabros M, Poirier B, Yousefi M, Cashman N

P139 Data-mining in PRO-ACT: diamonds in the rough
Sinani E, Walker J, Zack N, Katsovskiy I, Macklin E, Sherrman A

P140 Clinical profile of patients with amyotrophic lateral sclerosis followed at referral centers in Minas Gerais/Brazil
Prado L, Bicalho I, Lopes M, Gomez R, de Souza L, Teixeira A

P141 Motor neuron disease post poliomyelitis syndrome: Anthropometric profile in Brazilian patients
Castillo F, Piovesan R, Quadros A, Oliveira A

P142 Motor neuron disease/sequelae of poliomyelitis and post-poliomyelitis syndrome diagnoses in tertiary center
Quadros A, Motta M, Campos K, Munhoz C, Schmidt B, Oliveira A

P143 The clinical features of amyotrophic lateral sclerosis in Southwest China

P144 Survival and risk stratification of ALS patients – a population based study

P145 Exploring risk factors for ALS using the U.S. Medicare database
Freedman M, Wu J, Pfeffer R

P146 Distribution and persistence of the cytoskeletal protein BMAA: facilitating characteristics for human exposure
Purdie E, Metcalf J, Codd G

P147 A case-control geospatial analysis of residential environmental to environmental risk factors for ALS in New Hampshire and Vermont – cyanobacteria, agricultural chemicals, landfills and Superfund sites
Shi X, Torback NM, Guettet BG, Hanay J, Stommel EW, Andrew AS, Henegol PL, Callar TA, Bradley WG

P148 A geospatial analysis of the distribution of ALS cases in Florida and environmental risk factors – cyanobacteria, agricultural chemicals, landfills, and Superfund sites
Bradley WG, Shi X, Gaddipati N, Steele J, Guttet BN, Stommel EW, Andrew AS

P149 Environmental exposures as risk factors for ALS in the Lake Erie region of northern Ohio – the Cleveland clinic database and cyanobacteria
Pioro EP, Torback NM, Bullerjahn GS, Shi X, Andrew AS, Stommel EW, Bradley WG

P150 Aerosolization of cyanobacteria and cyanotoxins as risk factors for ALS/ neurodegeneration
Stommel EW, Cox PA, Bankac SA, Henegol PL, Grigel HB, Stommel AM, Hanay J, Torback NM, Muryl AB, Delong PA, Callar TA, Tsongalis GJ, Hickey WP, Gallagher TL, Davis DA, Bradley WG

P151 Bayesian modelling of potential association between soil mineral levels and small area spatial risk of ALS in Ireland

P152 Occupational exposure to lead, agricultural chemicals and electromagnetic fields and functional parameters at ALS diagnosis

P153 POSTER WITHDRAWN

P154 Environmental risk factors for ALS in NH and VT - a questionnaire-based case-control study
Andrew AS, Stommel EW, Duell EJ, Henegol PL, Tandan R, Callar TA, Bradley WG

P155 Retrospective analysis of heavy metal testing in ALS
Salomon K, Bertone V, Genge A, Massie R

P156 Population-based risks for cancer associated with ALS cases

P157 ApoE genotype and onset of cognitive impairment in ALS: no correlation. A population-based study

THEME 9
Genetics

P158 ALS Online Genetics Database, ALSoD: New features and current position
Abel O, Powell J, Andersen P; Al-Chalabi A

P159 Comprehensive genetic screening of 28 ALS-related genes in a Japanese ALS cohort

P160 Genetic analysis of cbl-c gene as a candidate gene for sporadic amyotrophic lateral sclerosis in African Americans
Nunez-Santana FL, Saeed M, Usacheva EA, Elliott IV D, Siddique NA, Siddique T

P161 Mutation screening in Brazilian ALS patients - an analysis of 7 genes in familial and sporadic cases
Beccari M, Moura Alves L, Mitne-Neto M, Zatz M

P162 NEK1 missense variants in 252 German and Nordic ALS families

P163 Fragment analysis identifies atypical C9ORF72 (G4C2) expansion patterns in familial and sporadic ALS/ALS/FTD in a North American population
Lowry J, Yan J, Kissely S, Siddique NA, Deng H-X, Siddique T

P164 C9ORF72 repeat expansion detection using short-read whole-genome sequencing data

P165 Update on C9ORF72 repeat expansion in Italian ALS patients
Mosca L, Talarini C, Lunetta C, Sansonna V, Penco S

P166 C9ORF72 epigenetic modifications in Italian amyotrophic lateral sclerosis patients
Calini D, Tiloca C, Verde F, Onesto E, Gentilini D, Ticozzi N, Silani V, Ratti A

P167 Clinical and genetic analysis or genotype-phenotype correlations with familial ALS of China Liu W, Xu X, Li X, Liu N, Deng M

P168 Genetic diagnosis of Chinese patients with amyotrophic lateral sclerosis using next generation sequencing
Zhang W, Zhijun L, Jingqiang T, Hongfu L, Wang N

P169 The G41D mutation in the superoxide dismutase 1 gene is associated with slow motor neuron progression and mild cognitive impairment in a Chinese family with amyotrophic lateral sclerosis
Jin Q


P171 Assessment of TREM2 rs75932628 association with amyotrophic lateral sclerosis in a Chinese population
Chen X, Chen Y, Wei Q, Shang H

P172 Two novel mutations of DCTN1 in Chinese sporadic amyotrophic lateral sclerosis patients Liu X, Yang L, Fan D

P173 TUB4A4 may not be a significant genetic factor in Chinese ALS patients
Li J, He J, Tang L, Xu J, Chen L, Ma Y, Zhang N, Fan D

P174 CHCHD10 was identified as ALS causative gene with complex mechanisms
Yan J, Ahmeti KB, Siddique NA, Ryan E, Lukas TJ, Deng H-X, Kissely S, Yong S, Ma Y, Miller N, Corbett N, Nicholson DA, Siddique T

P175 Ataxin 2 is not a disease modifier in a large series of ALS patients carrying the C9ORF72 expansion
P177 Analyses of the VCP gene in patients with sporadic amyotrophic lateral sclerosis, identify a novel mutation associated with increased susceptibility to oxidative stress


P178 Valosin-Containing Protein (VCP) gene analysis in a cohort of ALS patients: identification of a novel mutation

Tarafarini C, Mosca L, Maestri E, Caldarazzo Ienco E, Siciliano G, Lunetta C, Penco S

P179 A rapid functional decline type of sporadic ALS is linked to low expression of TTN


P180 Whole exome sequencing in sporadic ALS trios and familial ALS


P181 The role of de novo mutations in the development of sporadic ALS

van Doormaal PTC, Diekstra FF, van den Heuvel DMA, van Rheenen W, Overste D, Dekker AM, Schellevis RD, van Dammme F, de Bakker PIM, Francioli LC, Pasterkamp R, van den Berg LH, Veldink JD

P182 Identification of de novo mutations in sporadic amyotrophic lateral sclerosis


P183 De novo mutations of the FUS gene are the most frequent cause of sporadic ALS in very young onset patients

Hübbers A, Just W, Rosenhörm A, Muller K, Marroquin

P184 Subcellular localization and RNAs determine Fus architecture in different cellular compartments


P185 The phosphorylation of FUS by CK2 regulates its RNA-binding activity

Kuang L, Gal J, Chen J, Liu Y, Arenas A, Jia J, Zhou H

P186 The effects of steric hindrance on RNA editing using antisense oligonucleotides

Chaytow H, Popplewell L, Dickson G, Chen P

P187 Identification of novel substrates of the ALS-associated ribonuclease Angiogenin via RNA sequencing

Hogg M, Susdalezew S, Monsefi N, Prehn J

P188 Ubiquilin2 protein interacts with hnRNP family through PXX domain and hnRNP1 aggregates in sporadic ALS patients

Shi Y, Lukas T, Zhai H, Deng H-X, Siddique T

P189 New function of superoxide dismutase 1 in the nuclear compartment

Pansaraosa O, Gagliardi S, Dell’Orco M, Bordeaux M, La Salvia S, Maffioli E, Tedeschi GA, Ceredà C

P190 Genome-wide expression study identifies homeobox genes and transthyretin in C9orf72 expansion carriers


P191 C9orf72 repeat expansions dysregulate NETO1 in FTD-ALS

Porterfield V, Raymond M, Purow B, Foff E

P192 C9orf72 haploinsufficiency leads to compromised Rab29 mediated extracellular vesicle secretion, trans-Golgi trafficking and basal autophagy in C9orf72-associated ALS and FTD


P193 Mutations in hnrNPA2/B1 and hnrNPA1 linked to multisystem proteinopathy induce ER Stress and ER-Golgi trafficking defects reminiscent of ALS pathology

Sultana J, Shah D, Soo KY, Spencer D, Atkin J

P194 Defects in optineurin and myosin VI mediated intracellular and axonal trafficking in amyotrophic lateral sclerosis


P195 TLQP peptides in amyotrophic lateral sclerosis (ALS): animal model and human studies


P196 The novel ALS2-interacting small G protein Rab17 colocalizes with ALS2 in recycling endosomes

Ono S, Otomo A, Onodera W, Sato K, Mitsui S, Fukuda M, Hadano S

P197 ALS associated mutations in Matrin 3 alter vesicle secretion, trans-Golgi trafficking and basal autophagy in C9orf72-associated ALS and FTD

Rademacher S, Claus P

P198 Dissecting the interactome of RBM45 using immunoprecipitation and mass spectrometry


P199 The impact of Rho kinase (ROCK) and phosphatase and tensin homologue (PTEN) cross-talk on the neuronal cytokinetic in amyotrophic lateral sclerosis and spinal muscular atrophy

Radmacher S, Claus P

P200 Unraveling common mechanisms between manganese neurotoxicity and ALS-related motor neuron death

Obis T, Merwin S, Dziedzic J, Gonzalez K, Kariya S, TheMe 11

P201 BMAA Enantiomers in the central nervous system: Implications for neurodegenerative disease

Metcalf J, Lobner D, Banack S, Mash D, Cox G, Nunn P, Wyatt T, Cox P

P202 POSTER WITHDRAWN

P203 Prolonged intrathelial infusion of beta-N-methylamino-L-alanine (BMAA) induces preferential motor neuron damage and astrogliosis in the ventral horn of the spinal cord

Weiss J, Yin H

P204 Uptake and protein-association of the environmental neurotoxin BMAA (β-N-methylamino-L-alanine) in the neonatal central nervous system

Brittebo E, Karlsson O, Ersson L, Malmstrom T, Ilag L, Jiang L

P205 Locomotor and Electrophysiological Effects of an L-Beta-methylamino-alanine (L-BMAA) fed drosofila melangaster model of ALS-PDC

Goto J

P206 Chronic dietary exposure to BMAA in vervet results in BMAA brain concentrations similar to Guam ALS/PDC patients

Banack S, Metcalf J, Cox G, Powell J, Cox P

P207 BMAA in7duces proteotoxic stress in a novel in vitro ALS model

Dunlop RI, Cox P, Powell J

P208 Cyanobacterial toxins and their isomers in desert soils

Richer R, Banack S, Metcalf J, Cox P, Chatzielefthimia A

P209 The interaction of the environmental neurotoxin BMAA (Beta-Methylamino-L-Alanine) with mutant SOD1 in a zebrafish model of amyotrophic lateral sclerosis

Powers S, Lavin T, Kwon K, Sher R

P210 Neuropathologic examination of African Green Monkeys exposed to cyanobacterial toxin BMAA

Mash D, Davis D, Palmour R, Banack S, Bradley W, Cox P

P211 Proteomic characterization of cytoplasmic FUS aggregates

Kamelgarn M, Chen J, Zh H

P212 An emerging role of FUS at the neuromuscular junction: implications for ALS pathogenesis

So E, Mitchell JC, Vance C, Shaw CE

P213 Denervation of ALS neuromuscular junctions is caused by active zone loss and is ameliorated by exercise


P214 The effect of SOD1 zinc loss on the interaction with hCSC

Wright GA, Antonyuk SV, Hasnain SS

P215 Discrete unfolded species of SOD1 are revealed by native mass spectrometry

McAlary L, Yerbury J, Aquinila A

P216 Mutant SOD facilitates Nitrilation of Hsp90


P217 Regional- and lamina-specific alterations in Calretinin and NPY interneuron populations in the SOD1 mice and amyotrophic lateral sclerosis patients: A potential source for corticothalamic hyperexcitability

Clark R, Brizuela M, Blizzard C, King A, Young K, Dickson T

P218 Post-translational modifications promote formation of SOD1 oligomers with potential toxicity in ALS

Dokholyan N, Redler R, Proctor E, Fay J, Caplow M

P219 SOD1 transmission to motor neurons in a SOD1G85R mouse amyotrophic lateral sclerosis (ALS) model

Thomas EV, Fenton WA, Li D, Nagy M, Horwich AL, McGrath JM

P220 TDP-43 or FUS-induced misfolded wild-type SOD1 can spread intercellularly and induce SOD1 misfolding in recipient cells

Pokushevsky E, Grad L, Cashman N

P221 Spine loss is an early pathogenic event in the TDP-43 A315T mouse model of amyotrophic lateral sclerosis

13
P226 Determine the contribution from sense versus antisense transcript to C9orf72 hexanucleotide repeat expansion-mediated toxicity

P227 Bunina bodies in C9-ALS nervous systems
Stauffer J, Sabeni S, Schulte D, Ravits C

P228 ALS/FTD-associated SQSTM1 mutations exert their effects in a domain-dependent manner which may converge on autophagy

P229 Human spinal ventral horn contains a lower autophagic capacity than other CNS areas: Implication for selective vulnerability of motor system in ALS
Tokuda E, Andersen P, Bränntström T, Marklund S

P230 Disease-promoting immunological alterations in patients with amyotrophic lateral sclerosis

P231 Age-related changes in the inflammatory cytokine environment surrounding facial motoneurons
Katharesan V, Lewis M, Vink R, Johnson I

P232 Nav3, an axonal guidance protein aberrantly expressed in ALS
Bakkar N, Kousari A, Bowser R

THEME 12 In Vitro Experimental Models

P233 A new protocol for generating functional motor neurons from human stem cells
Cai Y, Guo X, Lavado A, Li Q, Akanda N, Martin C, Hickman J

P234 Banking and distribution of motor neurons derived from induced pluripotent stem cells: A focus on motor neuron diseases
Shelley B, Shue L, Mandefro B, Ho R, Ornelas L, Sareen D, Svendsen C

P235 C9orf72 iPSC-derived motor neurons and cortical neurons show features of ALS and FTD

P236 Generation and motor neuron differentiation of integration-free induced pluripotent stem cells (iPSCs) from ALS patients harboring FUS mutations
Guo W, Debray S, Patel S, Bohl D, Van Damme P, Verfaillie C

P237 Direct conversion of ALS patient fibroblasts harboring FUS mutations to induced neurons demonstrates FUS abnormalities in ALS neurons
Lim SM, Nahm M, Kim SH

P238 TDP-43 mislocalisation in primary motor and cortical neurons from a native-promotor-driven TDP-43 ALS mouse model as a high content automated image based screening platform

P239 Model of TDP-43-mediated cellular toxicity in human iPS-cell derived neurons

P240 A new cellular model of pathological TDP-43: the neurotoxicity of stably expressed CTFT25 of TDP-43 depends on the proteasome
Duan W, Liu Y, Li C

P241 Fibroblasts from patients with Amyotrophic Lateral Sclerosis (ALS) associated with mutations in TARDBP gene as model of TDP-43 proteinopathy
Hewamadduma C, Grieson A, Allen S, Higgsbottom A, Shaw PJ

P242 Disease model of amyotrophic lateral sclerosis/Parkinsonism-Dementia Complex in the Kii Peninsula

P243 An in vitro model for TBK1 haploinsufficiency in primary neuronal cultures

P244 Small molecule based high-efficiency differentiation of motor neurons from iPS cells derived from control and fALS patient fibroblasts
Scaber J, Muthiac R, Turner M, Cowley S, Talbot K

P245 Immortalized human neuronal progenitor cell line (ReNcell CX) is an ideal model to investigate regeneration of adult neurogenesis in the SVZ

P246 Development of personalized in vitro neuromuscular system for ALS/MND studies and drug screening

P247 Functional interrogation of active neuromuscular synapses between human stem cell-derived motoneurons and human skeletal muscle in vitro by a multi-electrode cantilever array

P248 NSC-34 motor neuron-like cells compared to motor neurons to investigate glutamate-mediated excitotoxicity

P249 Modulating the MCU/VDAC within the ERMC in ALS

P250 ALS models - the contribution of patients' lymphoblastoid cell cultures
Pansara O, Bordoni M, La Salvia S, Dell’Orco MA, Polveracce F, Diamanti L, Ceroni M, Gagliardi S, Cereda C

P251 Zebrafish C9orf72 loss of function models of amyotrophic lateral sclerosis
Rounding NP, De Vos KJ, Grierson AJ

P252 Identification of a novel neuroprotective drug for ALS using "ZNstress", a zebrafish high throughput phenotypic screen and validation in the sod1 G93A mouse model
McGown A, Binny C, Shaw PJ, Ramesh T

P253 Differential toxicity of nuclear RNA foci versus cytoplasmic localization of FTD/ALS-associated GGGGCC repeats in drosophila

P254 Generation and phenotyping of a novel mouse model of ALS-FUS using Bacterial Artificial Chromosome (BAC) technology

P255 In vivo investigation of truncated FUS loss- or gain-of-function
Valori C, Kerimoglou C, Skathivelu V, Naumann R, Fischer A, Neumann M

P256 A TDP-43Q331K transgenic mouse shows promise as a robust pre-clinical model of motor neuron disease (MND)
Stephenson J, Mead R, Jenkins T, Shaw PJ

P257 Absence of widespread mis-splicing in the pre-clinical phase of a native promotor-driven TDP-43 mouse model of ALS

P258 Stages of impaired eye movement control in amyotrophic lateral sclerosis are consistent with the model of sequential spreading of pTDP-43 pathology

P259 Mutant Profilin1 toxicity in the transgenic mouse model for ALS
Kiaei M, Basnakian A, Cozart M

P260 Further characterization of mutant Profilin1 transgenic mouse model for ALS
Cozart M, Basnakian A, Kiaei M

P261 Heterogeneity of Matrin-3 expression in the developing and aging murine central nervous system

P262 Role of CDNF in SOD1-G93A mouse model of amyotrophic lateral sclerosis
Voutilainen M, De Lorenzo F, Montonen E, Airavaara M, Tuominen R, Lindholm D, Saarma M

P263 Progranulin stimulates nerve regeneration and enhances functional recovery in vivo
Beel S, De Muynck L, Van Den Bosch L, Robberecht W, Van Damme P

P264 Prolonged relaxation phase as measured by twitch force in the SOD1 G93A mouse model
Sanchez B, Widrick JJ, Li J, Rutkove SB

P265 Respiratory plasticity in the SOD1(G93A) mouse model
Van Damme P

P266 Effect of gene copy number on dysphagia onset in SOD1-G93A transgenic mice
Tuominen R, Lindholm D, Saarma M
Therapeutic Strategies

P269 Effects of human mesenchymal stromal cells in ALS in vitro and in vivo models

P270 Rat mesenchymal stem cells modulate functional properties of primary cultured microglia via TGF-B secretion

P271 Autologous bone marrow mononuclear cell intrathecal transplantation may affect the survival duration in amyotrophic lateral sclerosis – Clinical study
Sane H, Sharma A, Paranjape A, Gokulchandran N, Saini JK, Badhe P

P272 Expansion of Treg by interleukin-2/antibody complexes slows disease progression in mutant SOD1 mice
Sheean R, McKay F, Cretney E, Nutt S, Scheller K, Perera JK, Badhe P

P273 Metabolic and immunologic effects of anti-interleukin-6 receptor antibody (MR16-1) in SOD1(G93A) mouse model of amyotrophic lateral sclerosis

P274 Astrocyte-derived TGF-B1 accelerates disease progression in ALS mice by interfering with neuroprotective functions of microglia and T cells
Endo F, Kominne O, Jin S, Watanabe SI, Matsuno M, Sobue G, Wyss-Coray T, Yamaoka K

P275 Ibudilast, a phosphodiesterase (PDe) 4 and PDe 3 inhibitor, demonstrates efficacy in two drosophila melanogaster models of ALS
Matsuda K, Makhay M, Iwaki Y

P276 Investigating upper motor neurons in amyotrophic lateral sclerosis

P277 The disulphide interchange activity and general chaperone function of Protein Disulphide Isomerase (PDI) are both protective against cell death pathways in amyotrophic lateral sclerosis (ALS)
Perri E, Parakh S, Thomas C, Spencer D, Atkin J

P278 POSTER WITHDRAWN

P279 Mahogunin ring finger 1 confers cytoprotection against mutant SOD1 and is defective in ALS mice

P280 Mutated SOD1 silencing in astrocytes: mechanisms leading to the protection of the neuromuscular function in amyotrophic lateral sclerosis
Rochat C, Aeberscher J, Bernard-Marissal Ne, Dirren E, Schneider B, Aeberscher P

P281 Immunotherapeutic targeting of monomer/ misfolded SOD1 in a canine disease model of ALS

P282 Trehalose decreases mutant SOD1 expression and alleviates motor deficiency in early but not end-stage amyotrophic lateral sclerosis in a SOD1- G93A mouse model
Li Y, Duan W, Li C

P283 Neuregulin 1 as a therapeutic target to modulate microglial activation in ALS
Song F, Liu J, Allender E, Ravits J, Loeb J

P284 Antisense oligonucleotides for SOD1 improves function and extends life of SOD1-G93A mice

P285 Genetic correction of C9orf72 repeat expansion mutation in ALS/FTD patient iPSCs

P286 A-4-methylpyridine induces activity and rescues hypoxiaceptable motor neurons from mutant FUS and SOD1 ALS patient-derived iPSCs

P287 Sigma 1 receptor activation modulates MER-associated crossstalk in the SOD1 model of amyotrophic lateral sclerosis

P288 Cell cycle inhibitor ameliorates motor neuron degeneration induced by polyglutamine-expanded androgen receptor

P289 Genotype specific impact of nuclear factor E2-related factor 2 (NRF2) treatment in animal models of amyotrophic lateral sclerosis
Nandar W, Neely E, Simmons Z, Connor J

P290 Systemic angiogenin delivery reverses defects in capillary density in SOD1G93A mice and extends lifespan in FUS (1-359) mice
O’Riordan S, Coughlan K, Halang M, Garcia C, Lewandowski S, Pehn J

P291 A high caloric diet leads to extended lifespan, motor dysfunction and lumbar spinal cord motor neuron loss in TDP-43A315T mice
Coughlan K, Halang M, Woods I, Hogg M, Pehn J

P292 MCT1 metabolic support in ALS disease pathogenesis
Philips T, Gaultier A, Rothstein J

P293 Targeting microtubules to improve outcomes in ALS
Clark J, Blizzard C, Chuckowree J, Lee C, Dickson T

P294 ADAMTS-4 is detrimental in a mouse model of amyotrophic lateral sclerosis

P295 Specific transduction of corticospinal motor neurons by AAV2 upon direct motor cortex injection
Jara J, Stanford M, Brooks C, Zhu Y, Devries S, Ozdinler H

P296 Motor neuronal targeting by self-complementary AAV9 via intra-CSF delivery in nonhuman primate for motor neurons diseases

P297 Chronic inhibitory effect of Riluzole on trophic factor production

P298 Ultra-high dose administration of methylcobalamin delayed symptomatic and neuropathological findings in wobbler mouse motor neuron disease
Ikeda K, Kaji R, Iwasaki Y

P299 Korea FDA approved Oral Yoo's solution (YS) as a mono-therapeutic agent that slows the progression of Amyotrophic lateral sclerosis (ALS) diseases
Yoo SH

P300 Tongue strength training accelerates tongue motility deficits in the SOD1-G93A rat model of amyotrophic lateral sclerosis
Ma D, Shuler J, Kumar A, Tungtir S, Nishimune H, Stanford J

P301 Preliminary investigation of safety and efficacy of fasudil in subjects with amyotrophic lateral sclerosis
Liu X, Fan D

P302 Long-term use of riluzole could improve the progression of sporadic amyotrophic lateral sclerosis patients in China
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P303 A Phase 1 trial of VM202 as a treatment for ALS: Safety data and a possible efficacy signal
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P305 The ALS Treatment Prize Challenge – leveraging the incentive prize model to accelerate preclinical drug development in ALS
Shinder S, Goodman J, Leitner M

P306 Connecting ALS patients and future clinical trials by TRICALS: A web-based international patient registration platform
van Eijk RPA, Veldink JH, van Es MA, van den Berg LH

P307 The ALS Untangled Table of Evidence
Bedlack R

THEME SW

Scientific Work in Progress

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Bedlack R
SW5 Corticospinal motor neuron development control genes as candidates for human ALS susceptibility


SW6 Onset of MND with isolated unilateral temporalis and masseter muscle wasting: Association with Arg521Cys FUS gene mutation

SW7 C9orf72 promoter hypermethylation is lost in reprogrammed ALS patient cells and re-acquired during motor neuron specification
Esanov R, Benatar M, Wahlstedt C, Zeier Z

SW8 ALS/FTD linked polyepitide produced by RAN translation of hexanucleotide repeat-containing RNAs encoded from the C9orf72 gene accumulate most in unaffected brain regions
Saberi S, Stauffer J, Schulte D, Baughn M, Ingersoll J, Al-Chalabi A

SW9 Increased phosphorylated neurofilament heavy (pNF-H) in CSF as a potential disease marker of canine degenerative myelopathy
Sibigroth C, Garcia V, Shaw G, Coates J, Garcia M

SW10 Increased in vivo glial activation in patients with primary lateral sclerosis (PLS) assessed with [11C]-PBR28 positron emission tomography

SW11 Ultra-high field magnetic resonance spectroscopy in ALS

SW12 Evaluating biomarkers in ALS efficacy trials
Macklin E, Schoenfeld D

SW13 New insights into novel prognostic biomarkers of longevity in ALS patients
Polo S, Atencio G, Calvo A, Juarez A, Cordero P, Martin MA, Moraleda JM, Martinez S, Perez E, Osta R, Garcia A

SW14 Quantification of CFS cytokines in ALS by a multiplexed bead-based immunoassay

SW15 Biochemical and clinical markers for motor neuron disease subtypes
Cavalli L, Dolciotti C, Ravaoli S, Paolicchi A, Rossi B, Bongioanni P

SW16 P2 receptor expression and modulation of the ERMCC in peripheral blood mononuclear cells from patients with amyotrophic lateral sclerosis

SW17 Modulating ER calcium uptake within the ERMCC in ALS
Malc A, Tadic V, Prell T, Goldhammer N, Stubendorff B, Witte OW, Grosskreutz J

SW18 A novel FUS knock-in mouse with frameshift mutation to investigate the pathobiology of FUS-ALS

SW19 A new FUS-A14 mouse model to dissect the pathobiology of FUS-ALS
Jaeger J, Park H, Acevedo-Arozena A, Fisher EMC, Devoy A

SW20 Exploring the functional properties of C9 and SOD1 patient specific iPSC-derived astrocytes and neurons
Rushton D, Thomesen G, Shelley B, Sances S, Manfredo B, Sareen D, Svendsen C

SW21 Extending survival in the murine model of ALS by promoting the M2 microglial state and enhancing neuronal trophic support
Snyder A, Neely E, Payne R, Simmons Z, Connor J

SW22 Motor neuron disease, glutamate excitotoxicity and oxidative stress: A pilot study in double blind with a dietary supplement donor curcumin

SW23 L-serine as a treatment for ALS
Miller RG, Levine TD, Cox PA, Bradley W-G

SW24 The CANALS study: A randomized, double-blind, placebo-controlled multicentre study to assess the efficacy on spasticity symptoms of a cannabidiol sativa extract in motor neuron disease patients

SW25 Lunasin Virtual Trial: A novel patient-centric design
Bedlack R

THEME CW

Clinical Work in Progress and Care Practice

CW1 MRI-histology correlates of cortical and white matter changes in post-mortem MND/FTD brain

CW2 Automated detection of fasciculations from 8-mode ultrasound images for motor neuron disease diagnosis
Bibbings K, Harding P, Combes N, Loram I, Hudson-Tole E

CW3 Nerve conduction studies in 154 cases of disease diagnosis
Bueno H, Schoenfeld D, Cudkowicz M, Atassi N, Rushton D, Mucumbira M, Gervais C, Leis A

CW4 Pain and ALS: A synergistic dichotomy?
Ren Y, Cui F, Yang F, Huang X

CW5 Benefits of scalp mobilization for improving range of motion and decrease pain of the amyotrophic lateral sclerosis shoulder
Gicalone A

CW6 Engagement in purposeful occupation comprising aerobic activity and muscular resistance has potential to improve function for people with MND
Carey H

CW7 Vital capacity—highly variable in patients with bulbar weakness?
Banno H, Schoenfeld D, Cudkowicz M, Atassi N

CW8 Diurnal intermittent abdominal pressure ventilation (IAPV) with pneumobelt in an ALS bulbar weakness?

CW9 Outcome studies of diaphragm pacing in ALS
Raheja N, Stephens HE, Walsh S, Lehman E, Morris A, Simmons Z

CW10 Uniform methods for clinical data collection: An epic approach for US ALS centers
Katz J, Sherman A, Walk D

CW11 Physical and cognitive fitness in young adult males and risk of amyotrophic lateral sclerosis at early age

CW12 The relationship between cognition and disease staging in ALS

CW13 A cognitive screening tool for ALS patients: The ALS-FTD-Cog, preliminary results
Beeldman E, Govaerts R, Raaphorst J, de Haan R, Schmand B, de Visser M

CW14 Longitudinal validation of the ALS-FTD-Questionnaire – preliminary results

CW15 Longitudinal assessment of frontal cognitive impairment in patients with motor neuron disease
Ferraro PM, Agosta F, Cau E, Riva N, Coperi M, Comi G, Filippi M

CW16 Profiling language impairment in ALS: Spelling

CW17 Speech performance in ALS: Influence of linguistic context and concomitant language impairment
Kuruvilla-Dudgale M, Green J, Hogan T, Custer C

CW18 Clinical interventions for swallowing and hyperactive gag reflex

CW19 The Straw Test for assessing bulbar dysfunction in ALS

CW20 Speech generating devices and young carers: A case study of twins caring for a parent with ALS
Kavanaugh MS, Banker-Horner L, Barkhaus P

CW21 Preserving legacy: A guide to message banking in one’s own voice
Costello J

CW22 Exploring individual quality of life in amyotrophic lateral sclerosis (ALS)
Mays I, Galvin M, Staines A, Hardiman O

CW23 Patient-centred decision support tools for ALS/MND

CW24 Online sharing and support among users of a MND forum
Bath PA, Ellis J

CW25 Level of satisfaction with assistive technology devices for patients with amyotrophic lateral sclerosis
Allegretti A, Clegg A

CW26 Different methods of wheelchair activation—single case study
Allegretti A, Clegg A

CW27 The implementation of a systematic screening regarding driving capacity in patients with ALS
Berry K, Bertone D, Genge A, Salmon K, Vitale A

CW28 Cervical spine disease mimicking bulbar ALS
Ross M, Gervais C, Leis A
# Programme of events/locations

## Friday 11 December

<table>
<thead>
<tr>
<th>Time</th>
<th>Event</th>
<th>Location</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>07.00 – 18.00</td>
<td>Registration International Symposium</td>
<td>Mediterranean Foyer</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>07.00 – 18.00</td>
<td>Speaker Room</td>
<td>Coquina North</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>09.00 – 10.15</td>
<td>Symposium Joint Opening Session</td>
<td>Mediterranean Salon 5-8</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>10.30 / 15.30</td>
<td>Refreshment breaks am/pm</td>
<td>Mediterranean Foyer</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>11.00 – 17.30</td>
<td>Symposium Scientific Sessions 2A/3A/4A</td>
<td>Mediterranean Salon 6-8</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>11.00 – 17.50</td>
<td>Symposium Clinical Sessions 2B/3B/4B</td>
<td>Mediterranean Salon 5</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>12.30 – 14.00</td>
<td>Lunch</td>
<td>Coquina South</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>18.00 – 20.00</td>
<td>Poster Session A</td>
<td>Coquina North</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>17.30 – 19.30</td>
<td>Project MinE (closed meeting)</td>
<td>Cordova 3</td>
<td>Lower Level</td>
</tr>
<tr>
<td>20.15 – 22.15</td>
<td>BMAA as an Environmental Trigger for Neurodegenerative Illness</td>
<td>Cordova 3</td>
<td>Lower Level</td>
</tr>
</tbody>
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## Saturday 12 December

<table>
<thead>
<tr>
<th>Time</th>
<th>Event</th>
<th>Location</th>
<th>Level</th>
</tr>
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<tbody>
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<td>Mediterranean Foyer</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>07.00 – 18.00</td>
<td>Speaker Room</td>
<td>Coquina North</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>08.30 – 17.40</td>
<td>Symposium Scientific Session 5A/6A/7A/8A</td>
<td>Mediterranean Salon 6-8</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>08.30 – 17.40</td>
<td>Symposium Clinical Session 5B/6B/7B/8B</td>
<td>Mediterranean Salon 5</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>10.00 / 15.30</td>
<td>Refreshment breaks am/pm</td>
<td>Mediterranean Foyer</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>12.30 – 14.00</td>
<td>Lunch</td>
<td>Coquina South</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>14.00 – 17.40</td>
<td>Symposium Alternative Sessions 7C/8C</td>
<td>Mediterranean Salon 1-2</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>18.00 – 20.00</td>
<td>Poster Session B</td>
<td>Coquina North</td>
<td>Lobby Level</td>
</tr>
</tbody>
</table>

## Sunday 13 December

<table>
<thead>
<tr>
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<th>Event</th>
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<th>Level</th>
</tr>
</thead>
<tbody>
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<td>Speaker Room</td>
<td>Coquina North</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>07.15 – 08.30</td>
<td>Western ALS Study Group (WALS)</td>
<td>Cordova 3</td>
<td>Lower Level</td>
</tr>
<tr>
<td>07.30 – 13.00</td>
<td>Registration International Symposium</td>
<td>Mediterranean Foyer</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>08.30 – 12.50</td>
<td>Symposium Scientific Sessions 9A/10A</td>
<td>Mediterranean Salon 6-8</td>
<td>Lobby Level</td>
</tr>
<tr>
<td>08.30 – 12.20</td>
<td>Symposium Clinical Sessions 9B/10B</td>
<td>Mediterranean Salon 5</td>
<td>Lobby Level</td>
</tr>
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<td>Lobby Level</td>
</tr>
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<td>Lobby Level</td>
</tr>
<tr>
<td>14.00 – 15.20</td>
<td>Symposium Joint Closing Session</td>
<td>Mediterranean Salon 5-8</td>
<td>Lobby Level</td>
</tr>
</tbody>
</table>
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