mnda motor neurone disease association

26th international symposium on ALS/MND

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Orlando USA

11 – 13 December 2015

Programme

Host: The ALS Association

ALS ASSOCIATION

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

mnda motor neurone disease association

26th international symposium on ALS/MND

Organiser of the symposium:

mnda motor neurone disease association

Motor Neurone Disease Association

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ALS ASSOCIATION

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The International Alliance of ALS/MND Associations

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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.

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 spirt of these meetings.

Together, we will end this disease.

Barbara J. Newhouse
President and CEO,
The ALS Association

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.

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

Location: Mediterranean 5-8

SESSION 1 JOINT OPENING SESSION

Chairs: *S Light (UK) K Talbot (UK)*

08.30 – 08.35 Welcome – *S Light (UK), K Talbot (UK)*

08.35 – 08.40 Welcome – *Barbara Newhouse, President and CEO, The ALS Association (USA)*

08.40 – 09.10 Addressing the burden of neurodegenerative disease: a global challenge – *R Shakir (UK)*

09.10 – 09.40 Is ALS a multistep process? – *N Pearce (UK)*

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

09.55 – 10.15 IPG Award and winner's research presentation

10.30 – 11.00 COFFEE Location: Mediterranean Foyer

Location: Mediterranean 6-8

SESSION 2A RNA BIOGENESIS AND PROCESSING

Chairs: *J Robertson (Canada) S McKnight (USA)*

11.00 – 11.30 The role of low complexity domains in proteins involved in ALS – *S McKnight (USA)*

11.30 – 11.50 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)*

11.50 – 12.10 Neuropathology in C9orf72-ALS is consistent with PURA sequestration and loss of function in motor neurons despite compensatory overexpression – *J Cooper-Knock (Sheffield)*

12.10 – 12.30 RBM45 homo-oligomerization mediates association with ALS-linked proteins and stress granules – *Y Li (USA)*

Location: Mediterranean 5

SESSION 2B CLINICAL MANAGEMENT

Chairs: *O Hardiman (Ireland) V Silani (Italy)*

11.00 – 11.30 Multidisciplinary care in ALS: measuring the immeasurable? – *O Hardiman (Ireland)*

11.30 – 11.50 The experimental treatment of bulbar symptoms in ALS with Nuedexta – *R Smith (USA)*

11.50 – 12.10 Treatment of medically refractory sialorrhea with electron beam radiotherapy (EBRT) to the parotid – *E Kasarskis (USA)*

12.10 – 12.30 A national study of muscle cramps in amyotrophic lateral sclerosis – *B Oskarsson (USA)*

12.30 – 14.00 LUNCH Location: Coquina South

Location: Mediterranean 6-8

SESSION 3A RNA LOCALIZATION AND DYSREGULATION

Chairs: *J Rothstein (USA) JP Taylor (USA)*

14.00 – 14.30 Altered RNA Metabolism in ALS and related diseases – *JP Taylor (USA)*

14.30 – 14.50 Nuclear pore abnormalities in C9orf72 ALS iPS neurons and tissue alter nucleocytoplasmic protein trafficking – *C Donnelly (USA)*

14.50 – 15.10 Characterization of C9orf72 expression in pathological cohort uncovers new clinical associations with specific C9orf72 transcripts – *M Van Blitterswijk (USA)*

15.10 – 15.30 In-depth analysis of sense and antisense RNA foci in a large pathological cohort of C9orf72 expansion carriers – *M DeJesus-Hernandez (USA)*

Location: Mediterranean 5

SESSION 3B HOLISTIC CARE

Chairs: *K Pearce (UK) J Armstrong (USA)*

14.00 – 14.30 Discussing prognosis with ALS/MND patients: Balancing hope and realism – *J Jacobsen (USA)*

14.30 – 14.50 Experiences of people with Motor Neurone Disease (MND) and their family caregivers receiving the news of diagnosis: A national survey – *S Aoun (Australia)*

14.50 – 15.10 The impact of family on decision-making in ALS care: The patient perspective – *G Foley (Ireland)*

15.10 – 15.30 Determinants of caregiver strain in carers of patients with ALS: A longitudinal study – *H Creemers (The Netherlands)*

15.30 – 16.00 COFFEE Location: Mediterranean Foyer

Location: Mediterranean 6-8

SESSION 4A DISEASE MODELS

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

16.00 – 16.30 Generating new mouse models of ALS – *C Lutz (USA)*

16.30 – 16.50 New mouse models of ALS show neuron survival and motor recovery after clearance of TDP-43 pathology – *A Walker (USA)*

16.50 – 17.10 A gain of toxicity by C9orf72 GGGGCC expansion in amyotrophic lateral sclerosis and frontotemporal dementia – *Q Zhu (USA)*

17.10 – 17.30 An *in vivo* model of BMAA-induced protein inclusions of Guam ALS/PDC – *P Cox (USA)*

Location: Mediterranean 5

SESSION 4B ALS HETEROGENEITY AND PROGRESSION

Chairs: *M Weber (Switzerland) J Katz (USA)*

16.00 – 16.30 Improving the classification of ALS – can we make it logical? – *A Al-Chalabi (UK)*

16.30 – 16.50 How common are ALS plateaus and reversals? – *R Bedlack (USA)*

16.50 – 17.10 Prediction of survival in individual ALS patients: Integration of clinical, cognitive, genetic and imaging data – *H Westeneng (The Netherlands)*

17.10 – 17.30 Prospective, longitudinal study comparing three outcomes measures – *P Andres (USA)*

17.30 – 17.50 Slow vital capacity and forced vital capacity in ALS: The same reality? – *S Pinto (Portugal)*

POSTER SESSION A

18.00 – 20.00
Location: Coquina North

18.00 – 18.20 Theme CW: Clinical work in progress and care practice

18.00 – 18.20 Theme 10: Molecular cell biology

18.20 – 18.40 Theme 6: Respiratory and nutritional management

18.20 – 18.40 Theme 9: Genetics

18.40 – 19.00 Theme 2: ALS heterogeneity and disease progression

18.40 – 19.00 Theme 13: *In vivo* experimental models

19.00 – 19.20 Theme 1: Clinical management

19.00 – 19.20 Theme 12: *In vitro* experimental models

Saturday 12 December 2015

Location: Mediterranean 6-8

SESSION 5A GENE THERAPY

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

08.30 – 09.00 Targeting RNA mis-splicing in SMA – *A Krainer (USA)*

09.00 – 09.30 Developing therapeutics for neuromuscular disease: from basic to translational studies – *B Kaspar (USA)*

09.30 – 09.50 A new AAV-based gene therapy approach for SOD1-linked ALS – *M G Biferi (France)*

Location: Mediterranean 5

SESSION 5B GENETIC COUNSELLING

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

08.30 – 09.00 ALS Genetics: Helping your patients and families understand – *N Siddique (USA)*

09.00 – 09.30 Genetic counselling in the post genomic world – a clinician's perspective – *C Shaw (UK)*

09.30 – 09.50 Pre-symptomatic ALS genetic counselling and testing: Experience and recommendations – *M Benatar (USA)*

10.00 – 10.30 COFFEE Location: Mediterranean Foyer

Location: Mediterranean 6-8

SESSION 6A CLINICAL-PATHOLOGICAL CORRELATES OF DISEASE PROGRESSION

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

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)*

Location: Mediterranean 5

SESSION 6B RESPIRATORY SUPPORT

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

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)*

12.30 – 14.00 LUNCH Location: Coquina South

Location: Mediterranean 6-8

SESSION 7A GENETICS AND EPIGENETICS

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

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)*

Location: Mediterranean 5

SESSION 7B NON-MOTOR SYMPTOMS OF ALS

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

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)*

Location: Mediterranean 1-2

SESSION 7C BIOMARKERS

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

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

Location: Mediterranean 6-8

SESSION 8A 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)*

Location: Mediterranean 5

SESSION 8B 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)*

Location: Mediterranean 1-2

SESSION 8C 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)*

POSTER SESSION B

18.00 – 20.00
Location: Coquina North

18.00 – 18.20 Theme 3: Biomarkers and outcome measures
18.00 – 18.20 Theme 8: Epidemiology
18.20 – 18.40 Theme 4: Imaging and electrophysiology
18.20 – 18.40 Theme 11: ALS pathogenesis and neurotoxicity
18.40 – 19.00 Theme 5: Cognitive change
18.40 – 19.00 Theme 14: Therapeutic strategies
19.00 – 19.20 Theme 7: Quality of life and palliative care
19.00 – 19.20 Theme SW: Scientific work in progress

Sunday 13 December 2015

Location: Mediterranean 6-8

SESSION 9A MOTOR NEURON VULNERABILITY

Chairs: *M Sendtner (Germany) S Burden (USA)*

08.30 – 09.00 Dysregulation of axonal RNA processing in motor neuron disease – *M Sendtner (Germany)*

09.00 – 09.30 Preserving neuromuscular synapses in ALS – *S Burden (USA)*

09.30 – 10.00 Connecting genes to pathways and networks in an effort to reveal the basis of selective motor neuron vulnerability – *H Ozdinler (USA)*

Location: Mediterranean 5

SESSION 9B CLINICAL TRIALS

Chairs: *A Ludolph (Germany) J Shefner (USA)*

08.30 – 09.00 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)*

09.00 – 09.20 Adaptive design single center phosphodiesterase type 4 (PDE4) inhibitor: Phase 1b/2a clinical trial – *B Brooks (USA)*

09.20 – 09.40 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)*

09.40 – 10.00 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)*

10.00 – 10.30 COFFEE Location: Mediterranean Foyer

Location: Mediterranean 6-8

SESSION 10A DISEASE MECHANISMS AND THERAPEUTIC TARGETS

Chairs: *B Turner (Australia) J Prehn (Ireland)*

10.30 – 10.50 Direct conversion as a method to subgroup ALS patient populations – *K Meyer (USA)*

10.50 – 11.10 A transcriptional comparison of human iPSC and mouse models of ALS defines the impact of motor neuron maturation, aging and disease – *R Ho (USA)*

11.10 – 11.30 Using C. Elegans to identify conserved modifiers of C9orf72-associated dipeptide toxicity – *T Lamitina (USA)*

11.30 – 11.50 *In vivo* chemiogenetics reveals neuroprotective excitation-related signalling in neurons and astrocytes in SOD1(G93A) mouse – *F Roselli (Germany)*

11.50 – 12.10 Bromodomain inhibitors regulate the C9orf72 locus in ALS – *Z Zeier (USA)*

12.10 – 12.30 Genetic removal of histone deacetylase 6 (Hdac6) delays the disease progression in a FUS mouse model of ALS – *E Pollari (Belgium)*

12.30 – 12.50 Protection by copper delivery in SOD-transgenic mice and the importance of the Copper Chaperone for SOD1 (CCS) – *J Beckman (USA)*

Location: Mediterranean 5

SESSION 10B COGNITIVE CHANGE

Chairs: *M Strong (Canada) M Hornberger (UK)*

10.30 – 11.00 Amyotrophic lateral sclerosis and frontotemporal dementia: understanding the spectrum – *M Strong (USA)*

11.00 – 11.20 A prospective population-based investigation of cross-modal emotional processing in ALS – *T Burke (Ireland)*

11.20 – 11.40 The social brain of patients with amyotrophic lateral sclerosis (ALS): the more the better – *H Aho-Oezhan (Germany)*

11.40 – 12.00 Relationship between cognitive and behavioural impairment and depression in a large ALS cohort – *J Rabkin (USA)*

12.00 – 12.20 Neuropsychiatric symptoms predictive of greater caregiver distress in ALS – *M Hornberger (UK)*

12.30 – 14.00 LUNCH Location: Coquina South

Location: Mediterranean 5-8

SESSION 11 JOINT CLOSING SESSION

Chairs: *K Talbot (UK) L Bruijn (USA)*

14.00 – 14.05 Poster Prize presentation

14.05 – 14.10 Invitation to Dublin 2016

14.10 – 14.40 Stem cell treatment strategies for neurodegenerative disease: from hype to hope? – *C Svendsen (USA)*

14.40 – 15.10 Patient 3.0: the centre of drug development – *B Muller (The Netherlands)*

15.10 – 15.20 Late breaking news

Poster sessions

THEME 1

Clinical Management

P1 Clinical diagnosis and multidisciplinary management in amyotrophic lateral sclerosis (ALS)
Santos Salvioni CC, Stanich P, Oda AI, 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
Couratier P, Marin B, Beghi E, Vial C, Bernard E, Lautrette G, Clavelou P, Guy N, Lemasson G, Rousso E, Debruxelles S, Cintas P, Arne-Bes M-C, Antoine J-C, Camdessanché J-P, Raymondeau M, Rudelle C, Nicol M, Logroscino G, Preux P-M

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
Aoun S, Breen L, Edis R, Oliver D, Harris R, Birks C, Henderson R, O'Connor M, Talman P

P5 Familial amyotrophic lateral sclerosis – what motivates first-degree relatives to participate in a preclinical study and how is the study process perceived?
Schaldecker C, Madinger M, Böhm S, Prudlo J, Weishaupt J, Ludolph A, Weydt P

P6 The spectrum of clinical opinion on genetic testing in ALS
Vajda A, McLaughlin RL, Thorpe O, Al-Chalabi A, Abrahams S, Hardiman O

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
Versterre S, Buus L

P10 Coin rotation task for assessing manual dexterity in ALS
Ortiz-Corredor F, Pena-Preciado M, Fernandez-Escobar L, Franco-Walteros J, Monroy-Medrano A, Mendoza-Pulido C

P11 Systematic investigation on needs and use of technological devices in Amyotrophic Lateral Sclerosis patients and caregivers: a nationwide, multicentre survey
Lunetta C, Andreazza C, Donvito G, Malberti I, Messina S, Russo M, Zuccarino R, Ravasi M, Pozzi S, Lizio A, Pompilio G

P12 Assistive equipment use by people with ALS/MND in Australia
Connors 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)
Sanjak M, Hirsch M, Habet N, Peindl R, Zheng N, Holsten S, Morgan B, Mash T, Englert D, Iyer S, Brooks BR

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
Takeuchi R, Toyoshima Y, Tada M, Shiga A, Miura T, Aoki K, Ikeuchi T, Nishizawa M, Kakita A, Takahashi H

P19 Tau oligomer accumulation in patients with globular glial tauopathy (GGT) type III
Toyoshima Y, Tanaka H, Takeuchi R, Shimizu H, Tada M, Shiga A, Yokota O, Kuroda S, Ikeuchi T, Kakita A, Takahashi H

P20 Elucidating the potential role of antecedent disease in amyotrophic lateral sclerosis
Mitchell C, Hollinger S, Goswami S, Polak M, Lee R, Okosun I, Glass J

P21 SOD1 mutation mimics a distal hereditary motor neuropathy
Siciliano G, Caldarazzo Ienco E, Lucchesi C, Fabbrini M, Rossi M, Bisordi C, Lo Gerfo A, Fogli A, Simi P

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, Ravaioli S, Paolicchi A, Rossi B, Bongioanni P

P23 The ALS Stratification Prize - using the power of big data and crowdsourcing for catalyzing breakthroughs in amyotrophic lateral sclerosis
Zach N, Küffner R, Alon H, Atassi N, Di Camillo B, Cudkowicz M, Garcia-Garcia J, Hardiman O, Leitner M, Li G, Mangravite L, Norel R, Norman T, Sherman A, Wang L, Stolovitzky G

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

P25 Correlation between progression rate, need for external aids and survival in patients with ALS
Díaz-Gómez 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
Rutter-Locher Z, Tewkesbury D, Stephenson L, Chio A, Ellis CM, Hardiman O, Leigh PN, Sidle KS, Veldink JH, Visser AE, van den Berg LH, Swingler R, Shaw CE, Pearce N, Al-Chalabi A

P28 Clinical characteristics of elderly patients with amyotrophic lateral sclerosis
Drory VE, Artman I, 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
Rafiq M, Lee E, Bradburn M, McDermott C, Shaw PJ

P31 Vitamin D: not protective in ALS
Blasco H, Madji Hounoum B, Dufour-Rainfray D, Patin F, Maillot F, Beltran S, Gordon PH, Andres C, Corcia P

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 M

P33 VALUES: The effects of low estrogen on executive functioning in a national sample of ALSci individuals suggests a neuroendocrine model of disease onset
Flaherty C, Brothers A, Kraft J, Harrison M, Legro R, Manni A, Yang C, Simmons Z

P34 Staging communication ability in advanced ALS patients
Wright AK, Langford VL, Holt EW, Bravver EK, Bockenek WL, Moore LA, Lucas NW, Smith NP, Nichols MN, Lindblom SS, Pacicco TJ, Holsten SE, Sanjak M, Ward AL, Fischer MP, Frumkin LH, Thorne SD, Oplinger H, Burgess CJ, Brooks BR

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
Blasco H, Garcon G, Patin F, Veyrat-Durebex C, Boyer J, Madji Hounoum B, Devos D, Vourc'h P, Andres C, Corcia P

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 Proteomic search for early biomarkers for amyotrophic lateral sclerosis
Muckova P, Wendler S, Rhode H, Prell T, Ringer TM, Stubendorff B, Hammer N, Schulz A, Witte OW, Grosskreutz J

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P41 The extracellular domain of p75 neurotrophin receptor is present in both urine and cerebrospinal fluid of people living with amyotrophic lateral sclerosis
Ferraiuolo L, Shepheard 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
Shepheard S, Wiklendt L, Schultz D, Tan V, Chataway T, Wuu J, Guillermin G, Benatar M, Rogers M-L

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 novel source of biomarkers in ALS
Gray E, Thompson A, Mager I, Talbot K, Andaloussi S, Wood M, Turner M

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, Rogaeva E, Tartaglia C, Zinman L, Robertson J

P47 Multi-frequency electrical impedance myography of the tongue: a biomarker for bulbar dysfunction?
McIlduff 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, Leitner M, Ferguson T, Cerarbaum J, Johns D

P49 Modeling EMPOWER data to improve the efficiency of ALS phase II/III clinical trials
Leitner ML, MacDougall J, Liu D, Ferguson TA, Cedarbaum J, Johns DR, Atassi N

P50 Serum creatinine is a liable 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
Hijikata Y, Katsuno M, Suzuki K, Hashizume A, Yamada S, Inagaki T, Banno H, Sobue G

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

P52 Longitudinal assessment of reachable workspace in ALS, using the Kinect system
Oskarsson B, Joyce N, de Bie E, Nicorici A, Kurillo G, Han J

P53 TGF-beta parallels Smad expression in ALS muscle and is a marker of disease progression
Si Y, Kim S, Cui X, Oh S, Anderson T, Al-Sharabati M, Kazamel M, King P

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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
Johannesen S, Kobor I, Grassinger J, Khomenko A, Baldaranov D, Blume J, Bruun T-H, Kassubek J, Müller HP, Ludolph A, Herr W, Schuierer G, Schulte-Mattler W, Prémont-Schwarz I, Schneider A, Deppe M, Bogdahn U

P55 Can an MRI-biomarker provide stabilization in G-CSF-treated ALS patients?
Baldaranov D, Khomenko A, Blume J, Kobor I, Johannesen S, Bruun T-H, Grassinger J, Herr W, Schuierer G, Schulte-Mattler W, Schneider A, Kassubek J, Deppe M, Bogdahn U

P56 Quality control of multicenter, high resolution T1 MRI as a brain biomarker in amyotrophic lateral sclerosis
Grosskreutz J, Dahnke R, Gaser C, Prell T, Agosta F, Bede P, Benatar M, de Carvalho M, Kalra S, Kassubek J, Reischauer C, Turner M, van Damme P, van den Berg LH, Weber M, Filippi M, NISALS quality control study group

P57 MRI correlates of (11C) PBR28 PET as a biomarker for ALS
Alshikho M, Zurcher N, Loggia M, Cernasov P, Yasek J, Akeju O, catana C, Rosen B, Cudkowicz M, Hooker J, Atassi N

P58 Increased functional connectivity within the sensorimotor resting-state network in ALS: a candidate MEG-based biomarker
Proudfoot M, Colclough GL, Menke RA, Talbot K, Woolrich MW, Nobre AC, Turner M

P59 The Canadian ALS Neuroimaging Consortium (CALSNIC)
Kalra S, Beaulieu C, Eurich D, Genge A, Hanstock C, Keith J, Korngut L, Lu J-Q, Shoesmith C, Yunusova Y, Zinman L

P60 A connectivity-based analysis of frontostriatal 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
Loewe K, Machts J, Kaufmann J, Petri S, Dengler R, Heinze H-J, Borgelt C, Vielhaber S, Schoenfeld MA

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 spatial statistics and voxel based morphometry study
Alruwaili A, Pannek K, Coulthard A, Henderson R, Kurniawan N, McCombe P

P65 Neuronal activation of behaviorally impaired patients with amyotrophic lateral sclerosis in tasks of executive functioning: A functional magnetic resonance imaging study
Keller J, Loose M, Böhm S, Aho-Özhan H, Gorges M, Pinkhardt EH, Kassubek J, Ludolph AC, Lulé D

P66 Post mortem brain imaging to interpret the in vivo MRI signature of MND
Menke R, Straathof M, Foxley S, Jbabdi S, Pallebage-Gamarallage M, Ansorge O, Miller K, Turner M

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
Machts J, Loewe K, Kaufmann J, Jakubiczka S, Abdulla S, Petri S, Dengler R, Heinze H-J, Vielhaber S, Schoenfeld MA, Bede P

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, Müller 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
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P73 Apparent diffusion coefficients distinguish amyotrophic lateral sclerosis from cervical spondylotic myelopathy
Koike Y, Kanazawa M, Terajima K, Watanabe K, Ohashi M, Endo N, Shimohata T, Nishizawa M

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, Sorarù G, Filippi M

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

P77 Power analysis of electroencephalographic mu rhythm in patients with ALS
Maležič T, Koritnik B, Zidar I, Berlot 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
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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
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Cognitive Change

P87 Structural brain correlates of cognitive and behavioural impairment in MND
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P88 Cortical atrophy correlates of cognitive and behavioral symptoms across the ALS-FTD continuum
Ratti E, Domoto-Reilly K, Caso C, Murphy A, Kelly K, Stepanovic M, Brickhouse M, Hochberg D, Cudkowicz M, Dickerson B

P89 18F-FDG-PET correlates of cognitive impairment in ALS
Calvo A, Canosa A, Cistaro A, Bertuzzo D, Fania P, Moglia C, Ilardi A, Cammarosano S, Pagani M, Chio A

P90 Association of oxidative stress biomarkers and cognitive event related potentials in patients with amyotrophic lateral sclerosis
Peña Sánchez M, Olivares Torres A, Zaldivar Vaillant T, Lara Fernández G, Riverón Forment G, González Quevedo A, Fariñas L, Pérez López C, Castillo Casañas Y, Martínez Bonne O, Cabrera Rivero A, Valdés Ramos L, Guerra Badía R, Oquendo Sánchez E, Fernández Carriera R, Menéndez Sainz MC

P91 Assessment of cognitive change for a person with ALS/MND within the clinic
Oliver D

P92 Longitudinal assessment of the Edinburgh Cognitive Assessment (ECAS) - Is there a learning effect in ALS patients?
Burkhardt C, Neuwirth C, Goldman B, Schneider U, Weber M

P93 Evaluation of the Edinburgh Cognitive and Behavioral ALS Screen (ECAS) in a US sample
Haines T, Lyter J, Walsh S, Morris A, Abrahams S, Simmons Z

P94 Development of parallel versions of the Edinburgh Cognitive and Behavioural ALS Screen (ECAS)
Crockford C, Kleynhans M, Wilton E, Al-Chalabi A, Hardiman O, Bak T, Abrahams S

P95 Cognitive continuum across amyotrophic lateral sclerosis (ALS) and frontotemporal dementia (FTD): an investigation on social cognition
de Souza L, Caramelli P, Prado L, Bicalho I, Guimarães H, Mioshi E, Bertoux M, Hornberger M, Teixeira AL

P96 Cognitive-behavioral changes in amyotrophic lateral sclerosis: Natural history and impact on patients and caregivers
Bock M, Duong Y-N, Kim A, Allen I, Murphy J, Lomen-Hoerth C

P97 Assessing the effect of a group therapy in parallel (SGP) for ALS patients and their caregivers
Riera Munt M, Carmona Rodriguez MJ, Morales Lorenzo N

P98 The Mismatch Negativity reveals subclinical cognitive dysfunction in ALS patients
Olivares Torres A, Iglesias Fuster J, Hernández Barros DM, Cuspineda Bravo E, Peña Sánchez M, Zaldívar Vaillant T, Lara Fernández G, Sánchez López J, Pérez Gesen C, del Río Bazán D, Gutiérrez Gil J, Beltrán León C, Machado Curbelo C

P99 Cognitive spectrum of Chinese patients with sporadic amyotrophic lateral sclerosis
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P100 Comparison of several respiratory assessments in ALS
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P101 The development of a multidisciplinary clinic to improve respiratory care of people with MND/ALS in Medway area
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P102 Worsening of respiratory function is predictive of prognosis of ALS even at a very early stage
Juntas-Morales R, Pageot N, Alphandery S, Camu W

P103 Multidisciplinary respiratory care support team improves respiratory management of inpatients with amyotrophic lateral sclerosis
Komai K, Tagami A, Ishida C, Takahashi K, Motozaki Y, Ikeda Y

P104 Air stacking effect on motor neuron disease / amyotrophic lateral sclerosis
Vital de Carvalho E, Holsapfel SG, Oda AL, Chieia MA, Oliveira ASB

P105 Average Volume Assured Pressure Support (AVAPS) in ALS patients with pure respiratory involvement: A randomized double-blind crossover trial
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P106 A decade of diaphragm pacing for ALS/MND: overall survival and current management of diaphragm pacing
Onders R, Katirji B, Elmo M, Kaplan C, Schilz R

P107 Diaphragm pacing might improve sleep in patients with amyotrophic lateral sclerosis
Ito H, Odake S, Kohriki S, Kawachi J, Kamei T, Onders R

P108 Analysis of function and survival in ALS patients with diaphragm pacing using virtual controls
Taylor A, Miller R, Onders R, Ennist D

P109 Factors associated with patient compliance with non-invasive ventilation in patients with ALS
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P110 Prolonged survival of non-invasive and invasive ventilation in Japanese patients with ALS
Kimura F, Hirose T, Nakamura Y, Shigekiyo T, Ishida S, Nakajima H

P111 Japanese neurologists recommendations to their patients and personal choices to tracheostomy with invasive mechanical ventilation (TIV) diverge but not to non-invasive mechanical ventilation (NIV)
Ogino Y, Ogino M

P112 Relationship between adverse clinical signs and progression of communication impairment in patients with amyotrophic lateral sclerosis on tracheostomy invasive ventilation
Nakayama Y, Shimizu T, Matsuda C, Haraguchi M, Mochizuki Y, Hayashi K, Hirai T, Nagao M, Kawata A, Oyanagi K

P113 Introduction of noninvasive ventilation and mechanically assisted coughing in patients with amyotrophic lateral sclerosis
Kano O, Hirayama T, Takazawa T, Ishikawa Y, Miura K, Yanagihashi M, Nagasawa J, Kyuzen M, Morioka H, Kawabe K, Ikeda K, Iwasaki Y

P114 Laryngeal response patterns to mechanical assisted cough in amyotrophic lateral sclerosis
Andersen T, Sandnes A, Brekka A-K, Hilland M, Clemm H, Fondenes O, Tysnes O-B, Heimdal J-H, Halvorsen T, Vollsæter M, Røksund OD

P115 Voluntary cough airflow predicts penetration/aspiration status in amyotrophic lateral sclerosis
Plowman E, Robison R, Tabor L, Randall S, Vu T, Gaziano J, Gooch C

P116 Relationship between evaluation of salivary volume and dysphagia in patients with motor neuron disease
Oda AL, Alves PCL, Silva R, Oliveira ASB

P117 Picc line as an alternative to gastrostomy in amyotrophic lateral sclerosis patients at advanced stages
Camu W, Pageot N, Juntas-Morales R, Alphandery S

P118 Arm-band evaluation of resting energy expenditure (REE) in ALS
Valeriani I, Zoni I, Bartolomei I, Andrini I, Fantoni E, Fiorito A, Salvi F

P119 Evaluating the potential of diet and food components as disease modifiers in amyotrophic lateral sclerosis (ALS)
Dawczynski CA, Ringer TM, Prell T, Stubendorff B, Witte OW, Lorkowski S, Grosskreutz J

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P120 Coping strategies used by people with motor neurone disease
Holland D, Mills R, Tennant A, Young C on behalf of the TONiC Group

P121 Psychological distress in motor neurone disease and its correlation with use of alcohol or drugs and other negative coping strategies
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P122 The fatigue to quality of life relationship in MND: Is gender moderating the effect?
Young C, Tennant A on behalf of the TONiC Group

P123 Existential loss in the context of motor neurone disease: A hermeneutic phenomenological study
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P124 The lost art of kissing
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P125 Teaching to take care: training course for home-based caregivers in amyotrophic lateral sclerosis: outcomes from a pilot project
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P126 Search of factors that affect the positive perception of ALS caregivers
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P127 Purpose in life and quality of life in ALS patient-caregiver dyads: A multilevel longitudinal analysis
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P128 The experience of burden over time in frontotemporal dementia and motor neuron disease
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P129 Caregiver burden in ALS – dimensions and difficulties
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P130 Clinical Research Continuum: Transparency, privacy and patient-centricity
Sherman A

P131 Research priorities in palliative and end of life care: MND results from a broad analysis of patient views
Cupid B, Van Godwin J, Tate T, Noble B, Smith R, Tuft J, Eley J, Black J, Stockton M, Cheesley A, Berry L, Loftus R, Dechamps A, Stevens E, Penny A, McEnhill L, McNair A, Fryett S, Kerr A, McIlfatrick S, Nelson A, Sivell S, Baillie J, Candy B, Best S, and Cowan K

P132 Transitional care for 100 patients with amyotrophic lateral sclerosis
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P133 The role of palliative care in a multi-disciplinary ALS clinic
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P134 Death with dignity in Washington and Oregon patients with amyotrophic lateral sclerosis
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P135 Update on the United States National Amyotrophic Lateral Sclerosis (ALS) Registry
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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, Antao 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, Sherman 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
Wei Q-Q, Chen X, Zheng Z-Z, Huang R, Guo X, Cao B, Zhao B, Shang H

P144 Survival and risk stratification of ALS patients – a population based study
Benoit M, Couratier P, Arcuti S, Copetti M, Fontana A, Nicol M, Raymondeau M, Logroscino G, Preux P-M

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

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

P147 A case-control geospatial analysis of residential exposure to environmental risk factors for ALS in New Hampshire and Vermont – cyanobacteria, agricultural chemicals, landfills and Superfund sites
Shi X, Torbick NM, Guetti BG, Haney J, Stommel EW, Andrew AS, Henegan PL, Caller 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, Guetti 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, Torbick 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, Banack SA, Henegan PL, Grigel HB, Stommel AM, Haney J, Torbick NM, Murby AL, Delong PA, Caller TA, Tsongalis GJ, Hickey WF, 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
Rooney J, Heverin M, Vajda A, Tobin K, Crampsie A, Staines A, Hardiman O

P152 Occupational exposure to lead, agricultural chemicals and electromagnetic fields and functional parameters at ALS diagnosis
Factor-Litvak P, Andrews L, Goetz R, Hupf J, Singleton J, Mitsumoto H, COSMOS Study Group ALS

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

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

P156 Population-based risks for cancer associated with ALS cases
Gibson S, Abbott DA, McLean H, Farnham J, Thai K, Figueroa K, Bromberg M, Pulst S-M, Cannon-Albright L

P157 ApoE genotype and onset of cognitive impairment in ALS: no correlation. A population-based study
Calvo A, Brunetti M, Barberis M, Restagno G, Manera U, Iazzolino B, Cammarosano S, Ilardi A, Canosa A, Moglia C, Chiò A

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Genetics

P158 ALS Online Genetics Database, ALSoD: New features and current position
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P159 Comprehensive genetic screening of 28 ALS-related genes in a Japanese ALS cohort
Nakamura R, Sone J, Atsuta N, Tohnai G, Watanabe H, Yokoi D, Nakatochi M, Watanabe H, Ito M, Katsuno M, Tanaka F, Hattori N, Izumi Y, Morita M, Taniguchi A, Abe K, Oda M, Mizoguchi K, Kaji R, Sobue G

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
Brenner D, Müller K, Wieland T, Weydt P, Hübers A, Meitinger T, Strom TM, Ludolph AC, Andersen PM, Weishaupt JH

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

P164 C9ORF72 repeat expansion detection using short-read whole-genome sequencing data
van Vugt JJFA, Shaw RJ, van Rheenen W, Dekker AM, Bekritsky M, Ajay SS, Shimul Chowdhury S, van Eijk KR, Schellevis R, van Es MA, Humbach T, Taft R, Bentley D, van den Berg LH, Eberle MA, Veldink JH

P165 Update on C9ORF72 repeat expansion in Italian ALS patients
Mosca L, Tarlarini C, Lunetta C, Sansona 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, Ju X, Li X, Liu N, Deng M

P168 Genetic diagnosis of Chinese patients with amyotrophic lateral sclerosis using next generation sequencing
Zhiying W, Zhijun L, Qingqing 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

P170 Optineurin mutations in patients with sporadic amyotrophic lateral sclerosis in China
Li C, Ji Y, Tang L, Chen L, Ye S, Liu X, Fan D

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 TUBA4A may not be a significant genetic factor in Chinese ALS patients
Li J, He J, Tang L, Xu L, 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 É, Lukas TJ, Deng H-X, Kinsley LM, Yong S, Yang Y, 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
Chio A, Conforti FL, Borghero G, Lunetta C, Penco S, Simone I, Mazzini L, Mora G, Caponnetto C, Mandrioli J, Monsurrò MR, Battistini S, Calvo A, Volanti P, Brunetti M, Barberis M, Mandich P, D'Alfonso S, Zollino M, Sabatelli M

P176 Exome sequencing reveals novel TBK1 mutation and rare TUBA4A variant in familial ALS

Elfasi A, Finch N, Wang X, DeJesus-Hernandez M, Brown P, Levitich D, Zuchner S, Asmann Y, Boylan K, Rademakers R, van Blitterswijk M

P177 Analyses of the VCP gene in patients with sporadic amyotrophic lateral sclerosis, identify a novel mutation associated with increased susceptibility to oxidative stress
Hirano M, Nishida M, Nakamura Y, Saigoh K, Sakamoto H, Ueno S, Isono C, Kusunoki S

P178 Valosing-Containing Protein (VCP) gene analysis in a cohort of ALS patients: identification of a novel mutation
Tarlarini 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
Atsuta N, Watanabe H, Hirakawa A, Nakamura R, Nakatochi M, Ishigaki S, Iida A, Ikegawa S, Katsuno M, Izumi Y, Morita M, Taniguchi A, Abe K, Mizoguchi K, Kano O, Imai T, Aoki M, Nakashima K, Kaji R, Sobue G

P180 Whole exome sequencing in sporadic ALS trios and familial ALS
Ahmeti K, Fecto F, Ahmeti K, Yang Y, Siddique NA, Yan J, Pericak-Vance M, Deng H-X, Ma Y, Siddique T

P181 The role of de novo mutations in the development of sporadic ALS
van Doormaal PTC, Diekstra FP, van den Heuvel DMA, van Rheenen W, Overste D, Dekker AM, Schellevis RD, van Damme P, de Bakker PIW, Francioli LC, Pasterkamp RJ, van den Berg LH, Veldink JH

P182 Identification of de novo mutations in sporadic amyotrophic lateral sclerosis
Ticozzi N, Calini D, Tiloca C, Kenna K, Verde F, Pensato V, Keagle P, Ratti A, Gellera C, Landers JE, Silani V

P183 De novo mutations of the FUS gene are the most frequent cause of sporadic ALS in very young onset patients
Hübers A, Just W, Rosenbohm A, Müller K, Marroquin N, Goebel I, Högel J, Thiele H, Altmüller J, Nürnberg P, Weishaupt JH, Kubisch C, Ludolph AC, Volk AE

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Molecular Cell Biology

P184 Subcellular localization and RNAs determine FUS architecture in different cellular compartments
Yang L, Zhang J, Kamelgarn M, Gal J, Zhu H

P185 The phosphorylation of FUS by CK2 regulates its RNA-binding activity
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P186 The effects of steric hindrance on RNA editing using antisense oligonucleotides
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P189 New function of superoxide dismutase 1 in the nuclear compartment
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P193 Mutations in hnRNPA2/B1 and hnRNPA1 linked to multisystem proteinopathy induce ER Stress and ER-Golgi trafficking defects reminiscent of ALS pathology
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P194 Defects in optineurin and myosin VI mediated intracellular and axonal trafficking in amyotrophic lateral sclerosis
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P195 TLQP peptides in amyotrophic lateral sclerosis (ALS): animal model and human studies
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P197 ALS associated mutations in Matrin 3 alter protein binding partners
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P206 Chronic dietary exposure to BMAA in vervets results in BMAA brain concentrations similar to Guam ALS/PDC patients
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P207 BMAA in7duces proteotoxic stress in a novel in vitro ALS model
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P210 Neuropathologic examination of African Green Monkeys exposed to cyanobacterial toxin BMAA
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P213 Denervation of ALS neuromuscular junctions is caused by active zone loss and is ameliorated by exercise
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P214 The effect of SOD1 zinc loss on the interaction with hCCS
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P215 Discreet unfolded species of SOD1 are revealed by native mass spectrometry
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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 cortical hyperexcitability
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P223 Depletion of TDP-43 induces mitochondrial fragmentation
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P224 pTDP-43 staging of amyotrophic lateral sclerosis in our 27 cases
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P225 A certain morphology of activated microglia, which correlates with TDP-43 pathology in ALS spinal cord
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P271 Autologous bone marrow mononuclear cell intrathecal transplantation may affect the survival duration in amyotrophic lateral sclerosis – Clinical study
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P272 Expansion of Tregs by interleukin-2/antibody complexes slows disease progression in mutant SOD1 mice
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P273 Metabolic and immunologic effects of anti-interleukin-6 receptor antibody (MR16-1) in SOD1(G93A) mouse model of amyotrophic lateral sclerosis
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P274 Astrocyte-derived TGF-β1 accelerates disease progression in ALS mice by interfering with neuroprotective functions of microglia and T cells
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P279 Mahogunin ring finger 1 confers cytoprotection against mutant SOD1 and is defective in ALS mice
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P281 Immunotherapeutic targeting of monomer/misfolded SOD1 in a canine disease model of ALS
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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
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P283 Neuregulin1 as a therapeutic target to modulate microglial activation in ALS
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P284 Antisense oligonucleotides for SOD1 improves function and extends life of SOD1-G93A mice
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P285 Genetic correction of C9orf72 repeat expansion mutation in ALS/FTD patient iPSCs
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P286 4-aminopyridine induces activity and rescues hypoexcitable motor neurons from mutant FUS and SOD1 ALS patient-derived iPSC
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P287 Sigma 1 receptor activation modulates ER-mitochondria crosstalk in the SOD1 model of amyotrophic lateral sclerosis
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P288 Cell cycle inhibitor ameliorates motor neuron degeneration induced by polyglutamine-expanded androgen receptor
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P289 Genotype specific impact of nuclear factor E2-related factor 2 (NRF2) treatment in animal models of amyotrophic lateral sclerosis
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P290 Systemic angiogenin delivery reverses defects in capillary density in SOD1G93A mice and extends lifespan in FUS (1-359) mice
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P291 A high caloric diet leads to extended lifespan, motor dysfunction and lumbar spinal cord motor neuron loss in TDP-43A315T mice
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P296 Motor neuronal targeting by self-complementary AAV9 via intra-CSF delivery in nonhuman primate for motor neurons diseases
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P300 Tongue strength training accelerates tongue motility deficits in the SOD1-G93A rat model of amyotrophic lateral sclerosis
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SW3 Enteroviral infection causes cytoplasmic aggregation and cleavage of TAR DNA binding protein-43
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SW4 Accrued somatic mutations trigger onset of amyotrophic lateral sclerosis: from hypothesis to mainstream thinking (2005-2015)
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SW5 Corticospinal motor neuron development control genes as candidates for human ALS susceptibility
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SW6 Onset of MND with isolated unilateral temporalis and masseter muscle wasting: Association with Arg521Cys FUS gene mutation
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SW8 ALS/FTD linked polydipeptides 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, Ravits J

SW9 Increased phosphorylated neurofilament heavy (pNF-H) in CSF as a potential disease marker of canine degenerative myelopathy
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SW10 Increased in vivo glial activation in patients with primary lateral sclerosis (PLS) assessed with [11C]-PBR28 positron emission tomography
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SW11 Ultra-high field magnetic resonance spectroscopy in ALS
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SW12 Evaluating biomarkers in ALS efficacy trials
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SW13 New insights into novel prognostic biomarkers of longevity in ALS patients
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SW22 Motor neuron disease, glutamate excitotoxicity and oxidative stress: A pilot study in double blind with a dietary supplement donor curcumin
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CW3 Nerve conduction studies in 154 cases of amyotrophic lateral sclerosis
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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 tracheostomized patient: A case report
De Mattia E, Iatomasi M, Garabelli B, Malberti I, Falcier E, Roma E, Rao F, Lunetta C, Sansone V

CW9 Outcome studies of diaphragm pacing in ALS
Raheja D, 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
Longinetti E, Mariosa D, Larsson H, Almqvist C, Lichtenstein P, Ye W, Fang F

CW12 The relationship between cognition and disease staging in ALS
Crockford C, Newton J, Elamin M, Stephenson L, Swingler R, Chandran S, Lonergan K, Pinto Grau M, Vajda A, Pender N, Chiwera T, Dalrymple L, Shaw C, Al-Chalabi A, Hardiman O, Abrahams S

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

CW14 Longitudinal validation of the ALS-FTD-Questionnaire – preliminary results
Govaarts R, Beeldman E, Beelen A, Grupstra H, van der Kooi A, de Haan R, Schmand B, Raaphorst J, de Visser M

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

CW16 Profiling language impairment in ALS: Spelling
Niven E, Newton J, Rewaj P, Colville S, Swingler R, Chandran S, Abrahams S, Bak T

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

CW18 Clinical interventions for swallow and hyperactive gag reflex
Armstrong J, Casey P, Veis S, Larsen K, Ajroud-Driss S, Heller S, Li J-M, Coleman J, Wolfe L, Sufit R, Siddique T

CW19 The Straw Test for assessing bulbar dysfunction in ALS
Ortiz-Corredor F, Pena-Preciado M, Franco-Walteros J, Fernandez-Escobar L, Monroy-Medrano A, Mendoza-Pulido C

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
Hogden A, Aho-Oezhan H, Loose M, Greenfield D, Ludolph A, Lulé D

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- a 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

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

Saturday 12 December

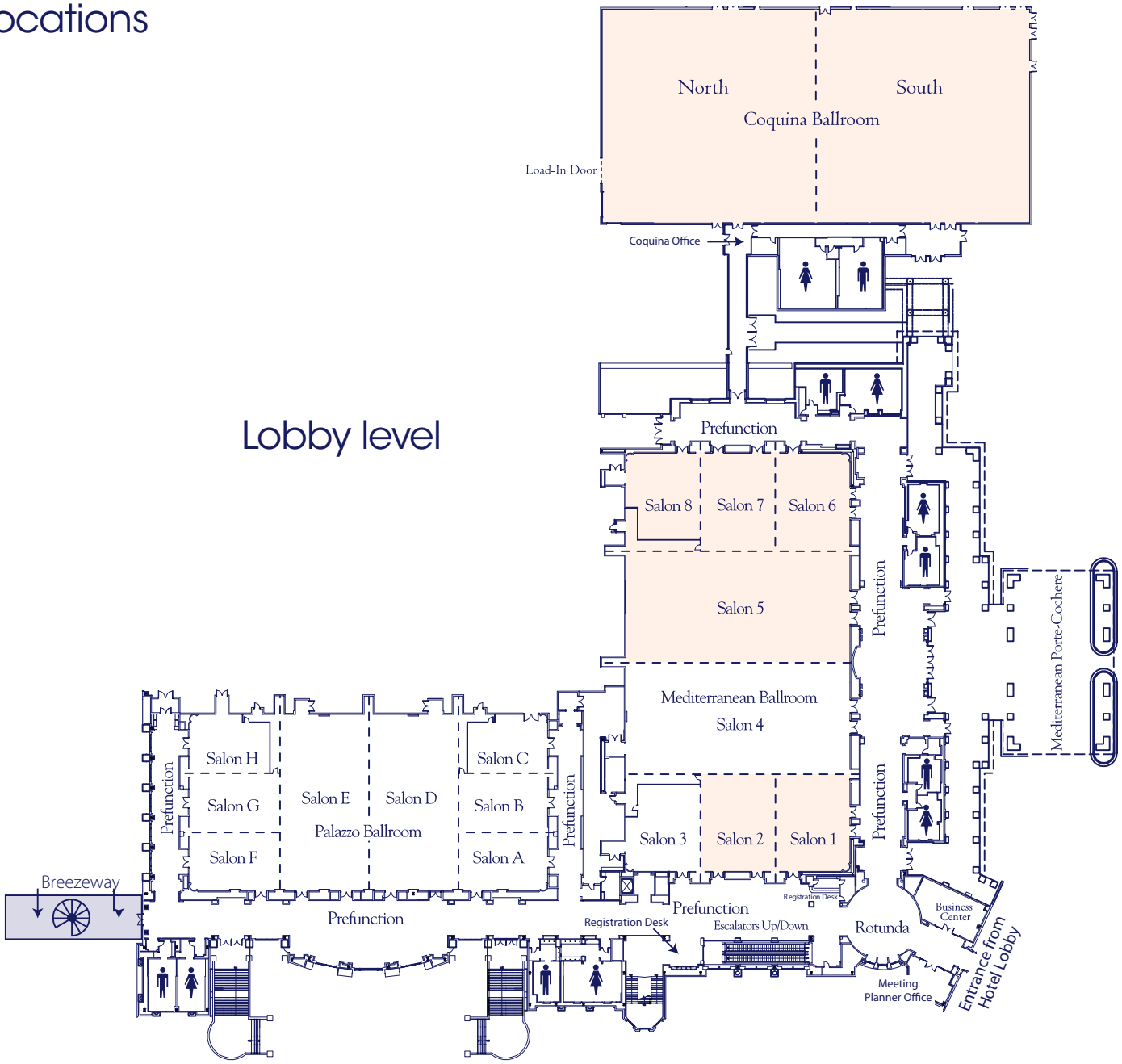
07.00 – 18.00	Registration International Symposium	*Mediterranean Foyer*	*Lobby Level*
07.00 – 18.00	Speaker Room	*Coquina North*	*Lobby Level*
08.30 – 17.40	Symposium Scientific Session 5A/6A/7A/8A	*Mediterranean Salon 6-8*	*Lobby Level*
08.30 – 17.40	Symposium Clinical Session 5B/6B/7B/8B	*Mediterranean Salon 5*	*Lobby Level*
10.00 / 15.30	Refreshment breaks am/pm	*Mediterranean Foyer*	*Lobby Level*
12.30 – 14.00	Lunch	*Coquina South*	*Lobby Level*
14.00 – 17.40	Symposium Alternative Sessions 7C/8C	*Mediterranean Salon 1-2*	*Lobby Level*
18.00 – 20.00	Poster Session B	*Coquina North*	*Lobby Level*

Sunday 13 December

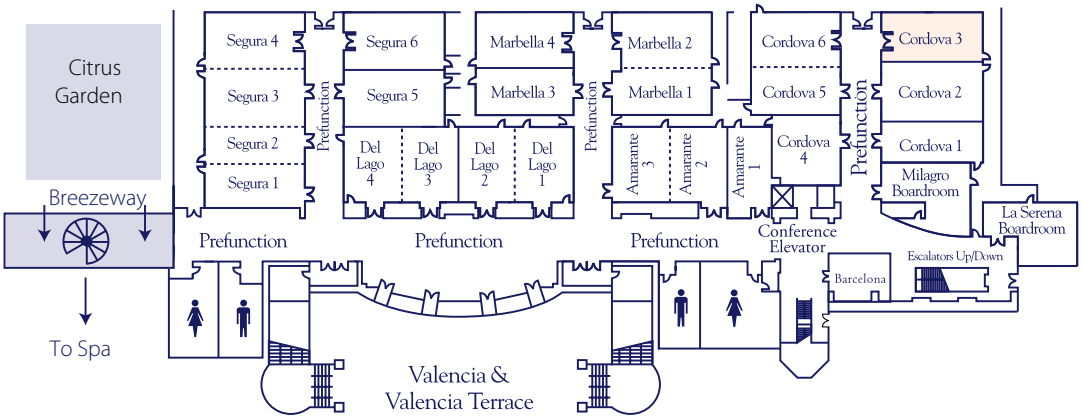
07.00 – 14.00	Speaker Room	*Coquina North*	*Lobby Level*
07.15 – 08.30	Western ALS Study Group (WALS)	*Cordova 3*	*Lower Level*
07.30 – 13.00	Registration International Symposium	*Mediterranean Foyer*	*Lobby Level*
08.30 – 12.50	Symposium Scientific Sessions 9A/10A	*Mediterranean Salon 6-8*	*Lobby Level*
08.30 – 12.20	Symposium Clinical Sessions 9B/10B	*Mediterranean Salon 5*	*Lobby Level*
10.00	Refreshment break	*Mediterranean Foyer*	*Lobby Level*
12.30 – 14.00	Lunch	*Coquina South*	*Lobby Level*
14.00 – 15.20	Symposium Joint Closing Session	*Mediterranean Salon 5-8*	*Lobby Level*

Locations

Lobby level



Lower level



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27 May 2016

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