

**mnda**

motor neurone disease  
association

25th international  
**symposium**  
on ALS/MND

5 – 7 December 2014

# Brussels

## BELGIUM

# Programme

**Host:** ALS Liga België vzw



**ALSLIGA.be**

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

## 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](mailto:symposium@mndassociation.org)

Website: [www.mndassociation.org](http://www.mndassociation.org)

## Host for the symposium:



**ALS LIGA.be**

### ALS Liga België vzw

Campus Sint Rafaël, Block H, 4th floor

Kapucijnenvoer 33 B/1,

3000 Leuven, Belgium

Tel: (-) 32 (0) 16 23 95 82

Fax: (-) 32 (0) 16 29 98 65

Email: [info@alsliga.be](mailto:info@alsliga.be)

Website: [www.alsliga.be](http://www.alsliga.be)

## Held in co-operation with:



### The International Alliance of ALS/MND Associations

Tel: (-) 1 215 568 2462

Fax: (-) 1 215 543 3366

Email: [alliance@als-mnd.org](mailto:alliance@als-mnd.org)

Website: [www.alsmndalliance.org](http://www.alsmndalliance.org)

## CME Accreditation

The 25th International Symposium on ALS/MND has been approved by the Federation of the Royal College of Physicians of the United Kingdom for 15 category 1 (external) CPD credit(s).



The '25th International Symposium on ALS/MND' is accredited by the European Accreditation Council for Continuing Medical Education (EACCME) to provide the following CME activity for medical specialists. The EACCME is an institution of the European Union of Medical Specialists (UEMS), [www.uems.net](http://www.uems.net).

The '25th International Symposium on ALS/MND' is designated for a maximum of 14 hours of European external CME credits. Each medical specialist should claim only those hours of credit that he/she actually spent in the educational activity.

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

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

## Welcome to Brussels!

ALS Liga Belgium is proud to host the 25th International Symposium on ALS/MND and the accompanying 22nd Annual Meeting of the International Alliance of ALS/MND Associations, for the first time being organized in Belgium.

Twenty-five years ago, the international ALS/MND community joined forces to gather annually and exchange the latest developments in science, care and support for the benefit of ALS/MND patients worldwide. Since then, major progress in all these areas has clearly been made, making us all proud. However, the ultimate goal – finding a cure for ALS/MND – has not yet been achieved, and a lot of effort is still ahead of us.

The 25th International Symposium on ALS/MND is also on the eve of the 20th anniversary of ALS Liga Belgium. Our patient organisation started small, as a self-help group by some Belgian ALS patients and their families. Today, we have built an organisation that deals with all aspects of support for ALS/MND patients, including communicating scientific progress on ALS/MND to our patients in a comprehensive way, accelerating scientific research on ALS/MND by continuous fundraising, lending equipment for mobility and communication, building awareness campaigns and interacting with national and regional policy makers and governmental agencies. We are delighted that, as a result, our not-for-profit organisation operates under the gracious patronage of Her Majesty the Queen of Belgium. For several years we have also, as a member of the International Alliance of ALS/MND Associations, taken responsibility for moving things forward in friendly collaboration with other countries.

Although Belgium is geographically rather small and even subdivided into multiple regions that speak different languages, it harbors a number of internationally-renowned opinion leaders in the field of ALS/MND research and care. We are thankful for the close collaboration with them that makes the 25th International Symposium on ALS/MND in Brussels possible.

I hope you all enjoy your time in Belgium and please contact us if you have any questions.

Hartelijk welkom, Soyez la bienvenue, Herzlich willkommen,

### **Evy Reviere**

CEO, ALS Liga Belgium

## Foreword

Welcome to the 25th International Symposium on ALS/MND. Recently, a wealth of new data on the genetics and biology of motor neuron degeneration has emerged. New avenues for the understanding of its mechanism have been opened. The speakers and presenters at the symposium will add novel data to these exciting new insights, and invited experts will critically address them. Increasing our knowledge of the biology of this disease will allow us to identify targets for intervention and opens perspectives for novel therapies and therapeutic approaches.

Providing optimal care for ALS/MND patients and their families is the main goal of ALS/MND associations and alliances. Critical evaluation of the treatments and approaches available is necessary to optimally contribute to the improvement of the quality of life of ALS patients and their caregivers. New developments in treatment of ALS patients, and novel ideas on how to approach ALS/MND and ALS/MND patients, will be presented.

This meeting hopes to contribute to a pathway to cure and care. The contribution from all of you is necessary for this. We are looking forward to a meeting that will bring us closer to our common goal.

### **Wim Robberecht**

Programme Committee Chair

## Friday 5 December 2014

Location: Gold Hall

### SESSION 1 **JOINT OPENING SESSION**

Chairs: *W Robberecht (Belgium) S Light (UK)*

**09.00 – 09.05** Welcome – *W Robberecht (Belgium) and S Light (UK)*

**09.05 – 09.20** Welcome – *Jo Vandeurzen, Minister of Welfare and Public Health (Belgium)*

**09.20 – 09.55** What is needed to advance a drug candidate into clinic trials? The perspective of one biotech company – *A Sandrock (USA)*

**09.55 – 10.05** International Alliance Humanitarian Award  
International Alliance Forbes Norris Award

**10.05 – 10.30** IPG Award and winner's research presentation

### 10.30 – 11.00 COFFEE

Location: Magritte Foyer

Location: Gold Hall

### SESSION 2A **RNA PROCESSING & DYSREGULATION**

Chairs: *C Shaw (UK) M Strong (USA)*

**11.00 – 11.30** Repeat associated non-ATG (RAN) translation in neurodegenerative disease – *L Ranum (USA)*

**11.30 – 11.50** Antisense and sense RNA foci derived from repeat expansions of C9orf72 have similar interactions but distinct expression patterns – *J Cooper-Knock (UK)*

**11.50 – 12.10** C9orf72 expression in amyotrophic lateral sclerosis and frontotemporal dementia – *L De Muynck (Belgium)*

**12.10 – 12.30** Hexanucleotide repeat expansions cause aberrant intron 1 retention in C9orf72 transcripts: an early event in the pathogenesis of C9ALS/FTD – *M Niblock (UK)*

Location: Copper Hall

### SESSION 2B **DIAGNOSIS/PROGNOSIS**

Chairs: *M Turner (UK) M Benatar (USA)*

**11.00 – 11.30** The challenge of early therapeutic intervention in ALS – *M Benatar (USA)*

**11.30 – 12.00** What does the study of premanifest disease contribute? Lessons from other neurodegenerative diseases – *S Tabrizi (UK)*

**12.00 – 12.20** Cortical excitability in familial C9orf72 ALS patients – *N Geevasinga (Australia)*

**12.20 – 12.40** Evaluation of routine laboratory tests as possible biomarkers of ALS in the preclinical and clinical phase – *V Drory (Israel)*

### 12.30 – 14.00 LUNCH

Location: Grand Hall 2

Location: Gold Hall

### SESSION 3A **PROTEIN MISFOLDING AND TOXICITY**

Chairs: *L Hayward (USA) V Buchman (UK)*

**14.00 – 14.30** The dynamics of protein folding: Pathologic aggregation in ALS mice follows test-tube behaviour – *M Oliveberg (Sweden)*

**14.30 – 14.50** Misfolded wild-type SOD1 induced by pathological FUS or TDP-43 transmits intercellularly and is propagated misfolding-competent – *E Pokrishevsky (Canada)*

**14.50 – 15.10** Co-expression of strain A- and B-aggregate forming human SOD1 mutants in mice: studies of aggregate structure and disease phenotype – *E Tokuda (Sweden)*

**15.10 – 15.30** Glutamate stimulates motor neurons to form intracellular p-TDP-43 aggregates – *N Shibata (Japan)*

Location: Copper Hall

### SESSION 3B **TRIALS AND TRIAL DESIGN**

Chairs: *M Cudkowicz (USA) PN Leigh (UK)*

**14.00 – 14.20** Does a placebo controlled clinical trial correctly estimate the treatment effect if the delivery method for the treatment and the placebo is potentially harmful? – *D Schoenfeld (USA)*

**14.20 – 14.40** Intracerebroventricular delivery of VEGF is feasible and safe in patients with amyotrophic lateral sclerosis, a phase I study – *P Van Damme (Belgium)*

**14.40 – 15.00** The effects of Tirasemtiv on measures of respiratory function in amyotrophic lateral sclerosis (ALS) – *J Andrews (USA)*

**15.00 – 15.20** Inflammation-associated plasma factors are associated with clinical response to NP001: a post hoc analysis of phase II clinical and laboratory data – *M McGrath (USA)*

### 15.30 – 16.00 COFFEE

Location: Magritte Foyer

Location: Gold Hall

## SESSION 4A AUTOPHAGY

Chairs: *J Atkin (Australia) S Hadano (Japan)*

- 16.00 – 16.20** P62/SQSTM1 deficiency accelerates motor neuron degeneration in SOD1H46R transgenic mice – *S Hadano (Japan)*
- 16.20 – 16.40** C9orf72 interacts with FIP200 and regulates the initiation of autophagy – *C Webster (UK)*
- 16.40 – 17.00** Increasing mTOR-independent autophagy enhances disease progression in a mouse model of ALS – *N Perera (Australia)*
- 17.00 – 17.20** Rab 1 rescues ER stress, macroautophagy and inhibition of ER-Golgi transport induced by mutant FUS in amyotrophic lateral sclerosis – *K Y Soo (Australia)*

Location: Copper Hall

## SESSION 4B ASSISTIVE TECHNOLOGY

Chairs: *S Petri (Germany) C McDermott (UK)*

- 16.00 – 16.30** AAC: from low to high tech – *A Reeves (UK)*
- 16.30 – 17.00** Use of brain computer interfaces in ALS – *A Kübler (Germany)*
- 17.00 – 17.20** Voice banking and voice reconstruction for MND patients – *P Rewaj (UK)*

## POSTER SESSION A

**17.30 – 19.30**

Location: Grand Hall 1

- 17.30 – 18.00** Theme 12A: Scientific work in progress
- 17.30 – 18.00** Theme 12B: Clinical work in progress and care practice
- 18.00 – 18.30** Theme 1: Improving diagnosis, prognosis and disease progression
- 18.00 – 18.30** Theme 6: Epidemiology
- 18.30 – 19.00** Theme 2: Imaging, electrophysiology and markers of disease progression
- 18.30 – 19.00** Theme 8: Human cell biology
- 19.00 – 19.30** Theme 3: Cognitive and psychological assessment and support
- 19.00 – 19.30** Theme 9: *In vitro* experimental models

## Saturday 6 December 2014

Location: Gold Hall

## SESSION 5A IN VITRO MODELLING

Chairs: *M Wiedau-Pazos (USA) K Eggan (USA)*

- 08.30 – 09.00** Are iPSCs living up to their promise? – *K Eggan (USA)*
- 09.00 – 09.20** A functional characterisation of C9orf72 iPSC-derived motor neurons – *R Mutihac (UK)*
- 09.20 – 09.40** The role of RBM45 in antioxidant responses in ALS – *N Bakkar (USA)*
- 09.40 – 10.00** Hexanucleotide repeat expansions in C9orf72 induce nucleolar stress and DNA damage in neuronal cell lines – *M Farg (Australia)*

Location: Copper Hall

## SESSION 5B CARE PRACTICE

Chairs: *H Mitsumoto (USA) T Meyer (USA)*

- 08.30 – 09.00** The role of eHealth in ALS: the Digital Agenda – *T Meyer (Germany)*
- 09.00 – 09.20** The impact of neck weakness and experiences of using neck orthoses in people with motor neurone disease – *C McDermott (UK)*
- 09.20 – 09.40** A description of pain in ALS – *Z Simmons (USA)*
- 09.40 – 10.00** Advance care planning in a Dutch tertiary ALS center, an evaluation of a Dutch care approach – *A Seeber (Netherlands)*

**10.00 – 10.30 COFFEE**

Location: Magritte Foyer

Location: Gold Hall

## SESSION 6A CELL BIOLOGY AND PATHOLOGY

Chairs: *P Shaw (UK) F Baralle (Italy)*

- 10.30 – 11.00** Functional and structural characterization of the hnRNP TDP 43 and its interactions – *F Baralle (Italy)*
- 11.00 – 11.20** Futsch/MAP1B is a translational target of TDP-43 and mitigates toxicity in motor neurons – *D Zarnescu (USA)*
- 11.20 – 11.40** Isoform-specific antibodies reveal region-dependent changes in C9orf72 protein levels in brains from ALS cases with repeat expansions in C9orf72 – *P McGoldrick (Canada)*
- 11.40 – 12.00** C9orf72 expansions are potentially pathogenic on three biological levels – *S Boeynaems (Belgium)*
- 12.00 – 12.20** RNA-dependent and RNA-independent aggregation of FUS in the cell cytoplasm: what structures become precursors of pathological inclusions in FUSopathies? – *V Buchman (UK)*
- 12.20 – 12.40** ELP3 as a disease modifier in ALS – *A Bento-Abreu (Belgium)*

Location: Copper Hall

## SESSION 6B EPIDEMIOLOGY

Chairs: *A Chio (Italy) L Cui (China)*

- 10.30 – 11.00** Genetics and phenotypes of amyotrophic lateral sclerosis in mainland China – *C Liying (China)*
- 11.00 – 11.20** Military service and amyotrophic lateral sclerosis in a population-based cohort – *M Weisskopf (USA)*
- 11.20 – 11.40** Association between premorbid diabetes mellitus and risk of amyotrophic lateral sclerosis in the Swedish population – *D Mariosa (Sweden)*
- 11.40 – 12.00** Head injury does not alter disease progression or neuropathologic outcomes in amyotrophic lateral sclerosis (ALS) – *C Fournier (USA)*
- 12.00 – 12.20** Predicting prognosis in ALS: A simple algorithm – *M Elamin (Ireland)*
- 12.20 – 12.40** Validation of a simple survival score for patients with ALS (ALS-SS) – *C Lunetta (Italy)*

## 12.30 – 14.00 LUNCH

Location: Grand Hall 2

Location: Gold Hall

## SESSION 7A GENETICS AND GENOMICS

Chairs: *J Veldink (Netherlands) G Nicholson (Australia)*

- 14.00 – 14.30** Finding ALS genes by means other than linkage – *J Hardy (UK)*
- 14.30 – 14.50** A novel locus at chromosome 1p associated with survival in patients with sporadic amyotrophic lateral sclerosis identified through an international genome wide meta-analysis – *I Fogh (UK)*
- 14.50 – 15.10** Genetic disease modifiers in individuals with C9orf72 repeat expansions – *M van Blitterswijk (USA)*
- 15.10 – 15.30** Exome sequencing identifies Matrin 3 as a new ALS gene – *J Johnson (USA)*

Location: Copper Hall

## SESSION 7B END OF LIFE DECISIONS

Chairs: *G Borasio (Italy) J Rosenfeld (USA)*

- 14.00 – 15.00** Assisted dying in ALS in the Netherlands and Belgium – *L van den Berg (Netherlands)*
- Assisted dying in ALS/MND: the Palliative Care viewpoint - *I Finlay (UK)*
- Assisted suicide and ALS: a 'middle way'? – *G Borasio (Switzerland)*
- 15.00 – 15.30** Discussion

Location: Silver Hall

## SESSION 7C BIOMARKERS (I)

Chairs: *A Ludolph (Germany) A Sherman (USA)*

- 14.00 – 14.15** Neurofilament light chain in blood is a prognostic, and a potential pharmacodynamic biomarker for amyotrophic lateral sclerosis – *A Malaspina (UK)*
- 14.15 – 14.30** CSF neurofilament light chain concentration reflects corticospinal tract microstructure in ALS – *M Turner (UK)*
- 14.30 – 14.50** Multi-center validation of a diagnostic assay for ALS – *R Bowser (USA)*
- 14.50 – 15.10** Evaluation of oxidative stress and other biomarkers at the baseline of a large ALS cohort study (ALS COSMOS) – *H Mitsumoto (USA)*
- 15.10 – 15.30** The role of albumin and creatinine in a population-based cohort of ALS patients – *A Calvo (Italy)*

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## 15.30 – 16.00 COFFEE

Location: Magritte Foyer

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Location: Gold Hall

### SESSION 8A MURINE MODELS

- Chairs: *L Greensmith (UK) B Turner (Australia)*
- 16.00 – 16.20** Development of mouse model for a newly discovered mutant profilin1 in fALS patients – *M Kiaei (USA)*
- 16.20 – 16.40** RNA processing alterations from ALS-linked mutations in FUS/TLS – *S Da Cruz (USA)*
- 16.40 – 17.00** Establishing a novel ALS knock-in mouse model with the ALS 8 mutation – *P Gaub (Canada)*
- 17.00 – 17.20** Physiological levels of gene expression in a BAC model of TDP-43-associated ALS lead to age dependent motor defects and cytoplasmic redistribution of TDP-43 – *D Gordon (UK)*
- 17.20 – 17.40** Transplant of light-sensitive stem cell-derived motor neurons to artificially restore muscle function in the SOD1G93A mouse model of ALS/MND – *B Bryson (UK)*

Location: Copper Hall

### SESSION 8B RESPIRATORY MANAGEMENT

- Chairs: *R Miller (USA) G Mora (Italy)*
- 16.00 – 16.20** Screening for respiratory failure in ALS using clinical questioning, respiratory function tests and transcutaneous carbon dioxide: which is the better tool? – *M Rafiq (UK)*
- 16.20 – 16.40** Independent factors associated with failed use of noninvasive ventilation (NIV) in patients with ALS/MND – *P Cazzolli (USA)*
- 16.40 – 17.00** Further analysis: diaphragm pacing in amyotrophic lateral sclerosis patients with chronic hypoventilation – *J Katz (USA)*
- 17.00 – 17.20** Palliative therapy during withdrawal of ventilation – a retrospective analysis of a 10 years experience in ALS – *T Meyer (Germany)*
- 17.20 – 17.40** Withdrawal of ventilation at the request of a patient with MND: exploring experiences of those involved – *C Faull (UK)*

Location: Silver Hall

### SESSION 8C BIOMARKERS (II)

- Chairs: *M Weber (Switzerland) M Kiernan (Australia)*
- 16.00 – 16.20** Phenotypic characterization and prediction of disease progression in ALS patients by metabolomics approach – *H Blasco (France)*
- 16.20 – 16.40** Smads as muscle biomarkers in amyotrophic lateral sclerosis – *P King (USA)*
- 16.40 – 17.00** Motor unit number index (MUNIX): ready for clinical ALS trials – a 15 months longitudinal multicentre trial – *C Neuwirth (Switzerland)*
- 17.00 – 17.20** Diagnostic utility of threshold tracking transcranial magnetic stimulation in ALS – STARD study – *N Geevasinga (Australia)*
- 17.20 – 17.40** Structural connectome analysis in ALS at multicenter level: a controlled study in 200 patients – *H Müller (Germany)*

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## POSTER SESSION B

### 17.45 – 19.30

Location: Grand Hall 1

- 17.45 – 18.20** Theme 4: Respiratory and nutritional management
- 17.45 – 18.20** Theme 10: *In vivo* experimental models
- 18.20 – 18.55** Theme 7: Genetics
- 18.20 – 18.55** Theme 11: Therapeutic strategies
- 18.55 – 19.30** Theme 5: Multidisciplinary care and quality of life

## Sunday 7 December 2014

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Location: Gold Hall

**SESSION 9A MODULATING SOD1 TOXICITY**

Chairs: *C Bendotti (Italy) H Durham (Canada)*

**08.30 – 08.50** Treatment for amyotrophic lateral sclerosis using AAV9 encoding a microRNA against SOD1 – *L Stoica (USA)*

**08.50 – 09.10** Novel in vivo active synthetic chemical chaperones as a new basis for ALS treatment – *A Gruzman (Israel)*

**09.10 – 09.30** Cu-ATSM: An effective treatment for high-expressing G93A-SOD1 mice expressing the human Copper Chaperone for SOD1 (CCS) – *J Beckman (USA)*

**09.30 – 09.50** Small molecules that block propagation of SOD1 misfolding in living cells – *L Grad (Canada)*

Location: Copper Hall

**SESSION 9B NUTRITIONAL ASSESSMENT AND MANAGEMENT**

Chairs: *V Drory (Israel) E Kasarskis (USA)*

**08.30 – 08.50** Nutrition and functional assessment in ALS patients – *C Gennings (USA)*

**08.50 – 09.10** A prospective multi-centre evaluation of gastrostomy in patients with MND – *T Stavroulakis (UK)*

**09.10 – 09.30** More on body mass index and survival in ALS – *S Rudnicki (USA)*

**09.30 – 09.50** Utility of self-report patient scales in the evaluation of dysphagia in individuals with ALS – *E Plowman (USA)*

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### 10.00 – 10.30 COFFEE

Location: Magritte Foyer

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Location: Gold Hall

**SESSION 10A NON NEURONAL CELLS**

Chairs: *L Van Den Bosch (Belgium) V H Perry (UK)*

**10.30 – 11.10** The contribution of local and systemic inflammation to neurodegeneration – *V H Perry (UK)*

**11.10 – 11.30** Contribution of CCL2/CCR2 axis in motor neuronal pathology of amyotrophic lateral sclerosis (ALS) – *C Bendotti (Italy)*

**11.30 – 11.50** Characterization of innate and adaptive immune responses in the hSOD1G93A-MCP1-CCR2 triple transgenic ALS mouse – *H Ozdinler (USA)*

**11.50 – 12.10** Increased in vivo glial activation in people with amyotrophic lateral sclerosis (ALS) – *N Atassi (USA)*

**12.10 – 12.30** Oligodendrocytes dysfunction in amyotrophic lateral sclerosis (ALS) – *J Leung (Australia)*

Location: Copper Hall

**SESSION 10B COGNITIVE CHANGE**

Chairs: *S Abrahams (UK) O Hardiman (Ireland)*

**10.30 – 11.00** Is ALS-FTD the same as FTD? – *J Snowden (UK)*

**11.00 – 11.20** The medical decision-making capacity in patients with amyotrophic lateral sclerosis – *R Spataro (Italy)*

**11.20 – 11.40** Longitudinal cognitive and behavioural screening in a large US cohort: Results from the COSMOS study group – *S Woolley (USA)*

**11.40 – 12.00** Multi-dimensional apathy in amyotrophic lateral sclerosis – *R Radakovic (UK)*

**12.00 – 12.20** Cognitive impairment and behavioural changes are associated with poor survival in ALS – *R Govaarts (Netherlands)*

**12.20 – 12.40** Prevalence, associations and course of depression in ALS: observations from a large cohort – *N Thakore (USA)*

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### 12.30 – 14.00 LUNCH

Location: Grand Hall 2

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Location: Gold Hall

**SESSION 11 JOINT CLOSING SESSION**

Chairs: *W Robberecht (Belgium) K Talbot (UK)*

**14.00 – 14.05** Invitation to Orlando 2015

**14.05 – 14.10** Poster Prize presentation

**14.10 – 14.20** Late breaking news

**14.20 – 15.00** Highlights of 2014  
Basic Research – *L Van Den Bosch (Belgium)*  
Clinical and Care Management Research – *J Ravits (USA)*



## Theme 1 Improving Diagnosis, Prognosis and Disease Progression

**P01 RED FLAGS FOR MUSCLE WEAKNESS: EXPLORING GP DECISION-MAKING AND REFERRAL PATHWAYS**  
MCDERMOTT CJ, BAXTER S, BEDFORD J

**P02 FOUNDATION OF THE NETHERLANDS ALS CENTRE IN 2003: REDUCING THE DIAGNOSTIC DELAY?**  
VISSER AE, SEELEN M, VELDINK JH, VAN DEN BERG LH

**P03 EXTENDING THE PHENOTYPE OF FOSMN SYNDROME: AN IMPORTANT ALS MIMIC**  
BROAD R, LEIGH PN

**P04 SPORADIC LOWER MOTOR NEURON DISEASE WITH A SNAKE EYES APPEARANCE ON THE CERVICAL ANTERIOR HORNS BY MRI: A NEW CLINICAL SUBTYPE?**  
SASAKI S

**P05 AMYOTROPHIC LATERAL SCLEROSIS (ALS) WITH LABORATORY ABNORMALITIES OF UNKNOWN SIGNIFICANCE (LAUS)] -- WHERE DOES IT BEGIN AND WHERE DOES IT END?**  
BROOKS BR, BRAVVER EK, DESAI UG, STORY SJ, LINDBLOM SC, PACICCO TJ, BOCKENEK WL, SANJAK MS, WARD AL, WRIGHT KA, LANGFORD VL, HOLSTEN SE, FISCHER MP, LARY C, NEMETH J, BELCHER SL, NICHOLS MS, SMITH NP, LUCAS NM, OPLINGER H

**P06 PARANEOPLASTIC SUBACUTE LOWER MOTOR NEURON SYNDROME ASSOCIATED WITH SOLID CANCER**  
VERSCHUEREN A, GALLARD J, ATTARIAN S, BOUCRAUT J, HONNORAT J, POUGET J

**P07 EXTRAPYRAMIDAL SYNDROME IN SPORADIC UPPER MOTOR NEURON-DOMINANT ALS WITH PURE TDP-43 PATHOLOGY**  
PRUDLO J, KASPER E, SCHUSTER C, KRAUSE BJ, BISKUP S, WALTER U, BENECKE R, BÜTTNER A, BESCHORNER R, NEUMANN M

**P08 ETOENCEPHALOGRAPHIC EVIDENCE OF CORTICAL MOTOR DYSFUNCTION IN ALS**  
PROUDFOOT M, ROHENKOHL G, GOULD I, TALBOT K, WOOLRICH M, NOBRE AC, WUJ J, BENATAR M, TURNER MR

**P09 PROGNOSTIC FACTORS ON THE COURSE OF FUNCTIONAL STATUS OF PATIENTS WITH ALS: A SYSTEMATIC REVIEW**  
CREEMERS H, GRUPSTRA H, NOLLET F, VAN DEN BERG LH, BEELEN A

**P10 ABSTRACT WITHDRAWN**

**P11 ALS ONSET AND PROPAGATION: INSIGHT FROM RESPIRATORY FUNCTION TEST**  
SOHN S-Y, KIM D-G, HONG Y-H, PARK K-H, SUNG J-J, LEE K-W, PARK K-S

**P12 THE RELATIONSHIP BETWEEN VOLUNTARY COUGH PRODUCTION AND SWALLOW SAFETY IN INDIVIDUALS WITH AMYOTROPHIC LATERAL SCLEROSIS**  
PLOWMAN EK, WATTS S, DOMER A, GAZIANO J, TABOR L

**P13 CLINICAL CORRELATION OF THE YAWNING REFLEX IN ALS PATIENTS**  
KWAN J, DIAZ M, ALTEMEMI N, ZILLIOX L, EMPENO R, RUSSELL J

**P14 AMYOTROPHIC LATERAL SCLEROSIS AND OXIDATIVE STRESS BIOMARKERS IN RELATION TO PHYSICAL EXERCISE**  
CHICO L, LOGERFO A, CALDARAZZO EI, PETROZZI L, ROCCHI A, CARLESI C, ORSUCCI D, SCHIRINZI E, SIMONCINI C, MANCUSO M, SICILIANO G

**P15 SIT TO STAND (STS) RATING SCALE: CONSTRUCT VALIDITY OF A NOVEL MEASURE OF LOWER EXTREMITY (LE) FUNCTION IN ALS PATIENTS**  
SANJAK M, OLDHAM J, BOYLES G, HOLSTEN SE, BOCKENEK W, STORY SJ, LINDBLOM SC, PACICCO T, BRAVVER E, RUSSO P, BROOKS BR

**P16 A DESCRIPTION OF PHYSICIAN PAIN MANAGEMENT PRACTICE IN ALS**  
SIMMONS Z, WALSH S, STEPHENS H

**P17 SELF-REPORTED MEASURES PREDICT SURVIVAL IN ALS**  
THAKORE N, PIORO E

**P18 BIOLOGICAL FOLLOW-UP IN AMYOTROPHIC LATERAL SCLEROSIS: CREATININE DECREASE AND FERRITIN INCREASE ARE PREDICTIVE OF A POOR PROGNOSIS**  
PATIN F, CORCIA P, MADJI HB, VEYRAT-DUREBEX C, RESPAUD E, PIVER E, BENZ DE BRETAGNE I, VOURCH P, ANDRES CR, BLASCO H

**P19 WHAT DO ALS PATIENTS DIE OF? – AN AUTOPSY STUDY OF 70 ALS PATIENTS**  
BURKHARDT C, NEURWIRTH C, SOMMACAL A, ANDERSEN P, WEBER M

**P20 MUSCLE ARCHITECTURE BY MEANS OF ECHOMYOGRAPHY, MULTIFREQUENCY ELECTRICAL IMPEDANCE MYOGRAPHY (MEIM) AND FORCE-TIME CURVE (F-TC) ANALYSIS IN ALS PATIENTS AS BIOMARKERS FOR PREDICTING MUSCLE DISEASE PROGRESSION**  
CORBIANCO S, BONGIOANNI P, TOSI C, D'ERAMO M, ROSSI B, DINI M

**P21 VIBRATION-INDUCED INHIBITION OF H-REFLEX IN ALS – A BIOMARKER FOR UPPER MOTOR NEURON DYSFUNCTION AND PREDICTOR OF FUNCTIONAL IMPAIRMENT**  
LEE MI, LANCE J, BURKE D, FITZPATRICK R, SIMON N, KIERNAN MC

## Theme 2 Imaging, Electrophysiology and Markers of Disease Progression

**P22 EVIDENCE FOR PERIPHERAL IMMUNE ACTIVATION IN AMYOTROPHIC LATERAL SCLEROSIS**  
CHEN X, GUO X, CHEN Y, SHANG H

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LIU X, GAO S, YU C, DENG M

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KO HK, ZHOU H, HAYWARD L

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NAGANO S, YAMAMOTO K, URUSHITANI M, FUJIWARA N, ARAKI T

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### P268 LIPOSOME-ENCAPSULATED H-FERRITIN IMPROVES SURVIVAL IN AN SOD1 MUTANT MOUSE MODEL OF ALS

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## P274 THE MND-ATTENUATING COMPOUND CUII (ATSM) ACTIVATES THE ANTIOXIDANT NRF2 PATHWAY IN CULTURED ASTROCYTES

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## P275 CORRECTING DEFECTIVE ENDOPLASMIC RETICULUM-MITOCHONDRIA INTERACTIONS AS A NEW THERAPEUTIC TARGET FOR ALS: CHARACTERISATION OF NOVEL DRUG SCREENS

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## P276 IMPROVEMENTS IN MOTOR FUNCTION, CA2+ CLEARANCE AND MARKERS OF ENDOPLASMIC RETICULUM STRESS WITH 6-GINGEROL TREATMENT IN SOD1 G93A ALS MICE

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## P277 CNS102 IMPROVES SURVIVAL AND MOTOR BEHAVIOR IN SOD1 MICE AND PROTECTS AGAINST EXCITOTOXICITY THROUGH MULTIPLE SIGNALING PATHWAYS

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## P281 IMPACT OF EXPIRATORY MUSCLE STRENGTH TRAINING ON BULBAR FUNCTION IN AMYOTROPHIC LATERAL SCLEROSIS: UPDATES FROM A RANDOMIZED SHAM-CONTROLLED CLINICAL TRIAL.

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## P282 MECHANICAL INSUFFLATION/EXSUFFLATION WITH HIGH FREQUENCY CHEST WALL OSCILLATION: RESULTS OF A CLINICAL TRIAL

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## P283 RELATIONSHIPS BETWEEN RILUZOLE AND TIRASEMTIV LEVELS ON OUTCOMES IN THE BENEFIT-ALS TRIAL

SHEFNER J, ANDREWS J, MENG L, BIAN A, WOLFF A, BENEFIT ALS STUDY GROUP

## P284 EFFECT OF RILUZOLE TREATMENT IN PATIENTS FROM EMILIA ROMAGNA, ITALY: A POPULATION BASED STUDY.

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## P285 DISCONTINUOUS RILUZOLE TREATMENT MAY PROVIDE A BETTER THERAPEUTIC ALTERNATIVE FOR ALS PATIENTS.

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## P286 FAST SKELETAL MUSCLE TROPONIN ACTIVATOR TIRASEMTIV INCREASES MUSCLE FUNCTION AND PERFORMANCE IN MOUSE MODELS OF SPINAL MUSCULAR ATROPHY

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## P287 DIPALS: PATIENT AND CARER EXPERIENCES OF DIAPHRAGM PACING IN MOTOR NEURON DISEASE

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## P288 PRO-ACT: EARLY RESULTS FROM THE LARGEST ALS CLINICAL TRIALS DATABASE

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### SW2 THE ROLE OF SOMATIC MUTATION IN SPORADIC AMYOTROPHIC LATERAL SCLEROSIS

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### SW3 GENOME-WIDE ASSOCIATION STUDY OF RARE VARIANTS IN ALS

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### SW4 MUTATION IN THE SENATAXIN GENE FOUND IN A CHINESE PATIENT AFFECTED BY LEFT LIMB WEAKNESS AND ATROPHY WITH JUVENILE ONSET

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### SW5 CROSS-DISEASE ANALYSIS OF COMMONLY DEREGULATED GENES AND PATHWAYS IN THE MOTONEURON DISEASES SPINAL MUSCULAR ATROPHY AND AMYOTROPHIC LATERAL SCLEROSIS

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### SW6 IMMUNOHISTOCHEMICAL ANALYSIS OF ERBB4 IN THE SPINAL CORD OF SPORADIC ALS PATIENTS

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### SW7 DEFECTIVE RECOGNITION OF ATG8/LC3 PROTEINS BY MUTANT P62/SQSTM1 IMPLICATES DYSREGULATION OF AUTOPHAGY IN ALS/FTLD

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### SW8 THE R399G MUTATION IN THE CYTOPLASMIC DYNEIN HEAVY CHAIN DISRUPTS GOLGI CISTERNAE IN HUMAN FIBROBLASTS.

GREEN R, ROSSOR A, SCOTO M, MUNTONI F, REILLY M, HAFEZPARAST M

### SW9 MAHOGUNIN RING FINGER PROTEIN 1, UBIQUITIN-PROTEIN LIGASE CONFERS NEUROPROTECTION AGAINST MISFOLDED PROTEIN AGGREGATION AND TOXICITY

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### SW10 DEVELOPMENT OF A CELLULAR TDP-43 AGGREGATION ASSAY BASED ON PROTEIN COMPLEMENTATION

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### SW11 GLUTATHIONE MONO-ETHYL ESTER REDUCES FORMATION OF CYTOSOLIC TDP-43 AGGREGATES IN NSC-34 CELLS

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### SW12 CLEARANCE OF THE ALS-ASSOCIATED TDP-43 PROTEINOPATHIES BY THE ACTIVATION OF HEAT SHOCK RESPONSE

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**SW14 CHARACTERISATION OF NEURONS DERIVED FROM INDUCED PLURIPOTENT STEM CELLS FROM MOTOR NEURONE DISEASE PATIENTS IDENTIFIES ALTERATIONS IN PROTEOSTATIC MECHANISMS**

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**SW15 EARLY DYSFUNCTION AND NON-CELL AUTONOMOUS DISEASE MECHANISMS IN A HUMAN IPSC- BASED MODEL OF ALS**

DEVLIN A-C, ZHAO C, BURR K, CHANDRAN S, MILES GB

**SW16 INVESTIGATING THE FUNCTIONAL CONSEQUENCES OF RNA PROCESSING DYSREGULATION IN ALS CELL MODELS**

DODD JE, SHAW PJ, HAUTBERGUE GM

**SW17 ESTABLISHING A STABLE CELL MODEL OF C9ORF72-ALS**

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**SW18 STUDYING GLIA-NEURONAL INTERACTION IN C9ORF72 EXPANSION MEDIATED ALS USING AN INDUCED PLURIPOTENT STEM CELL BASED IN VITRO MODEL**

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**SW19.5 IN VITRO ANALYSIS OF GLIAL CELL TOXICITY TO SPINAL CORD MOTOR NEURONS IN THE PRESYMPTOMATIC PHASE OF SOD1G93A ALS MOUSE MODEL - A POSSIBLE INVOLVEMENT OF TNFA, IL-6 AND NGF SIGNALLING.**

CHADI G, DUOBLES T, SCORISA JM, ALVES CJ, MAXIMINO JR

**SW20 EXTRACELLULAR AGGREGATED SOD1 IS TOXIC TO ASTROCYTES**

ROBERTS K, YERBURY J, CAMPBELL IL

**SW21 HSOD AND THE ALS ASSOCIATED MUTATION G93A INDUCE SPECIFIC LIPIDOMIC PROFILES IN CENTRAL NERVOUS SYSTEM IN TRANSGENIC MICE**

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**SW22 STEM CELL SURVIVAL IN THE SOD1 RAT MODEL OF ALS**

TURNER C, THOMPSON L

**SW23 CELLULAR THERAPY OF ANIMAL MODEL OF AMYOTROPHIC LATERAL SCLEROSIS BY TRANSPLANTATION OF HUMAN ASTROCYTES DERIVED FROM PLURIPOTENT STEM CELLS**

IZRAEL M, SLUTSKY G, GRANIT A, KRUSH L, MANZUR S, SKOROVSKY M, TAL Y, CHEBATH J, REVEL M

**SW24 NEW MOLECULAR THERAPY FOR AMYOTROPHIC LATERAL SCLEROSIS WITH SPINAL CORD TARGETING PEPTIDES**

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**SW25 CLINICAL TRIAL OF CEFTRIAXONE IN SUBJECTS WITH ALS - POST-HOC ANALYSIS USING CEFTRIAXONE CUMULATIVE DOSE**

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DOYLE L, JAGOE C

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**CW3 NUMBER OF C9ORF72 REPEAT EXPANSIONS AND PHENOTYPIC CORRELATION IN ALS**

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ZONDLER L, MÜLLER K, GROZDANOV V, BLIEDERHÄUSER C, WEYDT P, LUDOLPH AC, DANZER K, WEISHAUP T JH

**CW5 PROTEOMIC ANALYSIS OF MUSCLE TISSUE FROM PATIENTS WITH MOTORNEURON DISEASE AND CONTROLS**

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**CW5.5 CHOLESTEROL METABOLITES REGULATE MOTOR NEURON SURVIVAL VIA LIVER X RECEPTORS**

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**CW6 DEVELOPMENT OF A NEW PROTEIN BIOMARKER PANEL USING SELECTION REACTION MONITORING MASS SPECTROMETRY (SRM-MS)**

ZUBIRI I, WARD M, LIANG H-C, LEONI E, LU C-H, GREENSMITH L, PIKE I, MALASPINA A

**CW7 EVALUATION OF CORTICOSPINAL TRACT ALTERATION WITH DOUBLE INVERSION RECOVERY MAGNETIC RESONANCE IMAGING AS A NEW DIAGNOSTIC MARKER IN PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS**

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**CW7.5 ANATOMICAL CONNECTIVITY MAPPING AS A COMPLEMENTARY MEASURE TO FA IN ALS**

GABEL M, AL-CHALABI A, TSERMENTSSELI S, GOLDSTEIN L, GIULIETTI G, LEIGH PN, SIMMONS A, WILLIAMS S, CERCIGNANI M

**CW8 HIGH AND ULTRA-HIGH FIELD MR SPECTROSCOPY IN ALS**

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**CW9 EFFECTS OF THE ENDOCANNABINOID SYSTEM MODULATION ON EXCITOTOXIC STRESS OF RAT HYPOGLOSSAL MOTONEURONS**

SERZYNSKO-SOSNOWSKA M, NISTRU A

**CW10 THRESHOLD TRACKING REVEALS CHANGES OF PERIPHERAL AXONAL EXCITABILITY IN HEREDITARY SPASTIC PARAPLEGIA**

GUNKEL A, PRELL T, WITTE OW, GROSSKREUTZ J

**CW11 ACCELERATING INTERVENTION TO RESPIRATORY SUPPORT**

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**CW12 ALS AND AIRWAY CLEARANCE (ALSAC): IS THERE A BEST THERAPY FOR AIRWAY CLEARANCE IN PATIENTS WITH ALS?**

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**CW13 UPPER MOTOR NEURON DIAPHRAGM PARALYSIS IN AMYOTROPHIC LATERAL SCLEROSIS**

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**CW14 OUTCOMES OF AMYOTROPHIC LATERAL SCLEROSIS PATIENTS WITH PERCUTANEOUS ENDOSCOPIC GASTROSTOMY AND INVASIVE WENTILATION (TRACHEOSTOMY)**

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**CW15 SPEECH AND LANGUAGE THERAPY AS A RESOURCE IN THE CREATION OF PERSONAL LEGACY BOOKS**

DOYLE L

**CW16 TREATMENT OF NEUROGENIC STUTTERING IN MND: A CASE REPORT**

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**CW17 A PILOT STUDY ASSESSING A NEW EYE-WRITING DEVICE ALLOWING CURSIVE WRITING WITH SMOOTH PURSUIT EYE MOVEMENTS IN SUBJECTS WITH ALS**

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**CW18 PREVALENCE OF PERIPHERAL OEDEMA IN ALS AND CURRENT TREATMENT GUIDELINES**

SHAHBAZI M, PILOSSOPH L

**CW19 THE LOST ART OF KISSING**

SHAHBAZI M, CIANCI G, HOLZBERG S, LANGE D

**CW20 EXPLORATION OF CAREGIVING EXPERIENCES IN AMYOTROPHIC LATERAL SCLEROSIS: A PRELIMINARY ANALYSIS**

GALVIN M, CONNOLLY S, MAYS I, HARDIMAN O

**CW21 CAREGIVER BURDEN IN AMYOTROPHIC LATERAL SCLEROSIS (ALS): A PROSPECTIVE ANALYSIS**

GALVIN M, CONNOLLY S, MAYS I, HARDIMAN O

**CW22 PSYCHOLOGICAL PROBLEM OF CHILDREN WITH PARENTS WITH ALS**

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**CW23 ASSESSING PATIENT COGNITION AND BEHAVIOUR IN SPECIALISED ALS MULTIDISCIPLINARY CARE: A STUDY PROTOCOL**

HOGDEN A, CAGA J, GREENFIELD D, MIOSHI E

**CW24 APATHY, EMOTIONAL EXPRESSION AND PSYCHOLOGICAL ADAPTATION IN AMYOTROPHIC LATERAL SCLEROSIS**

BUNGENER C, DELGADILLO D, LEJEUNE S, SALACHAS F, PRADAT PF, BRUNETEAU G, LE FORESTIER N, LENGLET T, COURATIER P, LACOMBLEZ L

**CW25 CHALLENGES AND REWARDS OF SETTING UP A NETWORK MODEL: SOUTH WALES MOTOR NEURONE DISEASE (MND) CARE NETWORK**

GLEW R, HANCOCK K, JAMES J, DAWSON K, BAKER I, HADJIKOUTIS S

**CW26 REVERSING THE HUB AND SPOKE MODEL: PATIENT EXPERIENCES OF THE DEVELOPMENT OF A COMMUNITY BASED MULTI DISCIPLINARY MOTOR NEURONE DISEASE (MND) CLINIC TO MEET THE MOTOR NEURONE DISEASE ASSOCIATION 'STANDARDS OF CARE'**

ATKINSON C, BROWN C, BAKER M

**CW27 OCCUPATIONAL THERAPY- MAINTAINING FUNCTION THROUGH THE LIFE SPAN**

WARD A, HOLSTEN SE

**CW28 QUALITY OF LIFE IN AMYOTROPHIC LATERAL SCLEROSIS: A REVIEW**

MAYS I, GALVIN M, CONNOLLY S, HARDIMAN O

**CW29 MISCONCEPTIONS IN PALLIATIVE CARE FOR PATIENTS WITH ALS**

KAPLAN L, SHAHBAZI M, KASSEBAUM N, LANGE D, PILOSSOPH L

**CW30 COLLABORATION BETWEEN ALS AND PALLIATIVE SPECIALISTS IN DENMARK**

JAKOBSEN S, PUJIC SN, GREDAL O



## Friday 5 December

<b>07.00 – 18.00</b>	Registration International Symposium	<i>Registration Lobby</i>	<i>Level -1</i>
<b>07.00 – 18.00</b>	Speaker Room	<i>Registration Lobby</i>	<i>Level -1</i>
<b>07.15 – 08.15</b>	NEALS	<i>Studio 211/212</i>	<i>Level 2</i>
<b>09.00 – 10.30</b>	Symposium Joint Opening Session	<i>Gold Hall</i>	<i>Level 0</i>
<b>10.30 / 15.30</b>	Refreshment breaks am/pm	<i>Magritte Foyer</i>	<i>Level 0</i>
<b>11.00 – 17.20</b>	Symposium Scientific Sessions 2A/3A/4A	<i>Gold Hall</i>	<i>Level 0</i>
<b>11.00 – 17.20</b>	Symposium Clinical Sessions 2B/3B/4B	<i>Copper Hall</i>	<i>Level 0</i>
<b>12.30 – 14.00</b>	Lunch	<i>Grand Hall 2</i>	<i>Level -2</i>
<b>17.30 – 19.30</b>	Poster Session A	<i>Grand Hall 1</i>	<i>Level -2</i>
<b>17.30 – 19.30</b>	Project MinE (closed meeting)	<i>Studio 201 A/B</i>	<i>Level 2</i>

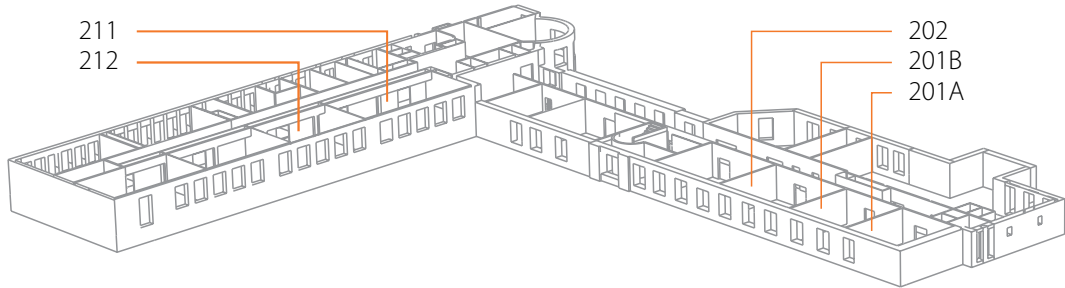
## Saturday 6 December

<b>07.00 – 18.00</b>	Registration International Symposium	<i>Registration Lobby</i>	<i>Level -1</i>
<b>07.00 – 18.00</b>	Speaker Room	<i>Registration Lobby</i>	<i>Level -1</i>
<b>08.30 – 17.40</b>	Symposium Scientific Sessions 5A/6A/7A/8A	<i>Gold Hall</i>	<i>Level 0</i>
<b>08.30 – 17.40</b>	Symposium Clinical Sessions 5B/6B/7B/8B	<i>Copper Hall</i>	<i>Level 0</i>
<b>14.00 – 17.40</b>	Symposium Alternative Sessions 7C/8C	<i>Silver Hall</i>	<i>Level 0</i>
<b>10.00 / 15.30</b>	Refreshment breaks am/pm	<i>Magritte Foyer</i>	<i>Level 0</i>
<b>12.30 – 14.00</b>	Lunch	<i>Grand Hall 2</i>	<i>Level -2</i>
<b>17.30 – 19.00</b>	Cochrane Neuromuscular Group	<i>Studio 201 A/B</i>	<i>Level 2</i>
<b>17.45 – 19.30</b>	Poster Session B	<i>Grand Hall 1</i>	<i>Level -2</i>

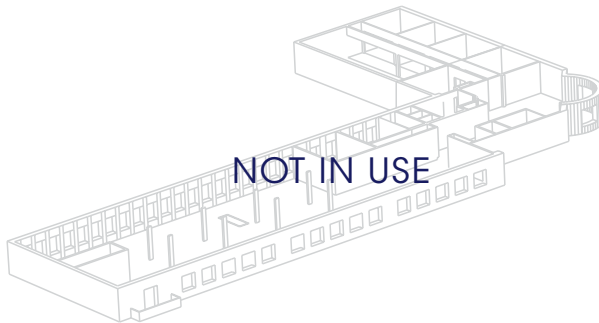
## Sunday 7 December

<b>07.00 – 08.30</b>	WALS	<i>Studio 201 A/B</i>	<i>Level 2</i>
<b>07.00 – 14.00</b>	Speaker Room	<i>Registration Lobby</i>	<i>Level -1</i>
<b>07.30 – 13.00</b>	Registration International Symposium	<i>Registration Lobby</i>	<i>Level -1</i>
<b>08.30 – 12.30</b>	Symposium Scientific Sessions 9A/10A	<i>Gold Hall</i>	<i>Level 0</i>
<b>08.30 – 12.40</b>	Symposium Clinical Sessions 9B/10B	<i>Copper Hall</i>	<i>Level 0</i>
<b>10.00</b>	Refreshment break	<i>Magritte Foyer</i>	<i>Level 0</i>
<b>12.30 – 14.00</b>	Lunch	<i>Grand Hall 2</i>	<i>Level -2</i>
<b>14.00 – 15.00</b>	Symposium Joint Closing Session	<i>Gold Hall</i>	<i>Level 0</i>

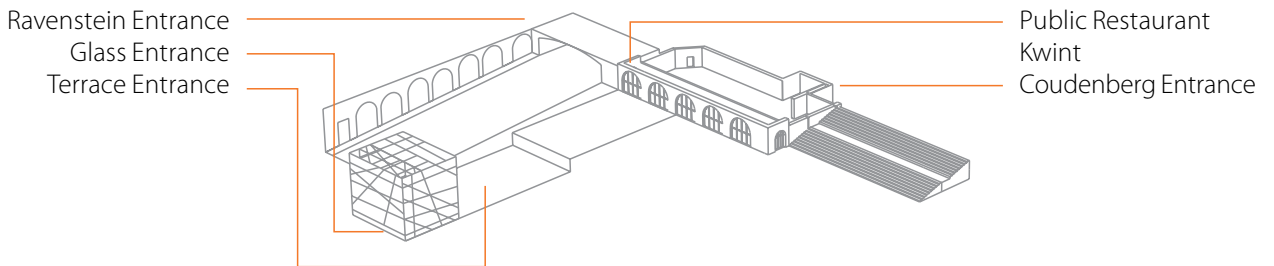
## Level 2



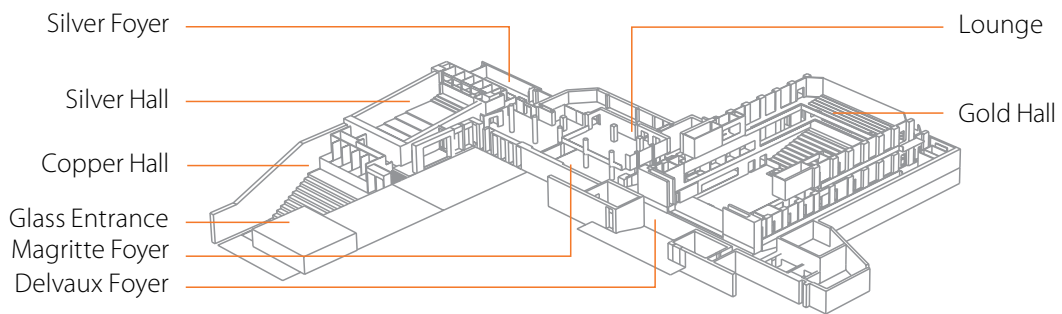
## Level 1



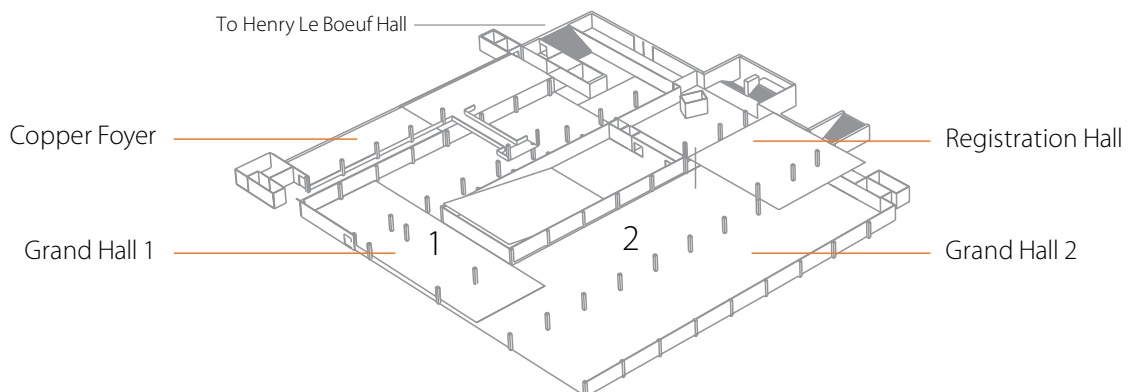
## Entrances



## Level 0



## Level -2 and -1



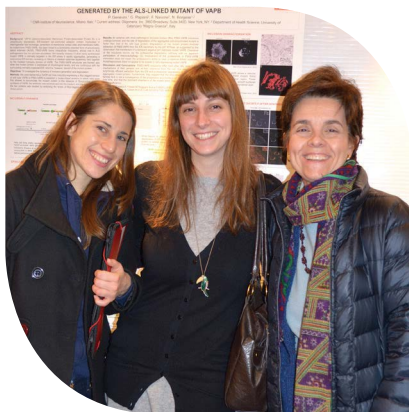
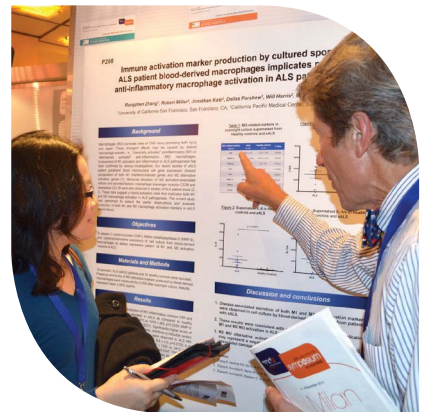
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For further information please contact  
**Motor Neurone Disease Association**  
PO Box 246 Northampton NN1 2PR

Tel: +44 (0) 1604 611 845  
Fax: +44 (0) 1604 624 726  
Email: [symposium@mndassociation.org](mailto:symposium@mndassociation.org)  
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