ENCALS

European Network to Cure ALS

ENCALS meeting 2023

Barcelona, Spain • 11th–14th July

Scientific Programme https://www.encals.eu/meetings/barcelona/

ENCALS

European Network to Cure ALS

Contents

Welcome to BARCELONA!

Dear attendees of the ENCALS

It is an honor and a privilege to warmly welcome you to ENCALS that will take place in the beautiful city of Barcelona. On behalf of the entire organizing committee, I would like to express our sincere gratitude for joining us in this scientifically significant event.

This congress represents a unique opportunity for researchers, academics, and professionals from around the world to share knowledge, ideas, and advancements in their respective fields. Barcelona, renowned for its rich history, impressive architecture, and vibrant cultural atmosphere, will be the perfect setting for this high-level scientific exchange.

During the days of the congress, we are thrilled to offer you a packed agenda of keynote speeches, roundtable discussions, and presentations of cutting-edge research. Our goal is to foster an environment conducive to dialogue and collaboration, where lasting connections can be established with colleagues and experts from various areas.

Additionally, Barcelona offers a wide range of opportunities to explore and enjoy during your stay. From the marvelous Mediterranean beaches to the architectural treasures of Gaudí, this captivating city will provide you with unforgettable experiences and moments of relaxation after an intense scientific journey.

We would like to express our gratitude to our sponsors and collaborators for their unwavering support, as this congress would not be possible without them. Furthermore, I want to extend my appreciation to all members of the organizing committee for their dedication and hard work in the planning and coordination of this event.

We hope you make the most of your participation in the congress, engaging in stimulating discussions, establishing new connections and collaborations, and taking away valuable ideas and knowledge to enrich your own research and professional careers.

On behalf of the organizing committee, we extend our warmest welcome to Barcelona and wish you a pleasant and successful stay at the ENCALS. We hope this event will be a turning point in your professional trajectory, and that you return home with new discoveries and lasting friendships.

See you soon in Barcelona!

Mònica Povedano Padanes
Bellvitge University Hospital (Barcelona)

Information for Presenters

POSTER DETAILS:

- Velcro stickers will be available at the registration desk.
- You have to bring your poster with you (printed).
- All posters must to be designed on A0 portrait size.
- There are two poster sessions. Every poster needs to be presented during both poster sessions (it is ok if someone else presents the next day, as long as someone is present at the poster).

TIMING PRESENTATIONS

- Plenary keynote speakers have 20 minutes + 5 minutes for questions.
- Abstract presenters have 8 minutes + 2 minutes for questions.
- Rapid fire presenters have 2 minutes + 1 minute for changing speaker. No questions. All Rapid fire presenters have a poster as well, so if people have questions, they can find the rapid fire presenter there.

TECHNICAL PRESENTATION DETAILS PRESENTERS:

- There is a speaker room available; here you can upload your presentation. This room is available from Monday 10-7, 10 AM. You can upload your presentation at any day of the event.
- If you prefer, you can also send your presentation to the technical staff in advance. Please email your presentation to envioppt@gmail.com.
  - Make sure to name the file (and mention it in the email) with your complete name, day and time of presentation, session and title.
  - To be sure, please bring your presentation with you on a USB stick as well.
  - If you have emailed your presentation, please always check before your presentation if it came through correctly.
- Please make sure your PowerPoint presentation is in format 16:9.

GENERAL INFORMATION

LOCAL ORGANISER
Dr. Mònica Povedano Panades
Bellvitge University Hospital (Barcelona)

VENUE
CCIB – Centre de Convencions Internacional de Barcelona
Plaça de Willy Brandt, 11-14
08019 Barcelona
https://ccib.es/en/

CONTACT INFORMATION
General ENCALS office
Abstract submission, programme, and remaining questions.

Contact person:
Danique van der Gaauw (ENCALS Office manager)
E: info@encals.eu

ENCALS Barcelona 2023 – local organising team
Sponsorship, general meeting information

Contact person:
Maria Unzué (congress & event manager Torres Pardo)
E: m.unzue@torrespardo.com
www.torrespardo.com

Accomodation requests or group booking
Contact: Accommodation team ENCALS
E-mail: encals@bnetwork.com
Telf: +34 93 550 03 50

REGISTRATION
https://www.emma.events/encals2023

Contact person:
Carolina Rovira
c.rovira@torrespardo.com

Each ENCALS centre is entitled to one free registration for a PhD student. Please email name, centre affiliation and dietary requirement of this PhD student to info@encals.eu.
Deadline for this registration is on July 1, 2023.

NORMAL REGISTRATION – UNTIL APRIL 30, 2023, AT 5 PM (CEST)

- Post graduate students – €150
  Young researchers who are still in training (students).
- Established researchers (including post docs) – €270
  Everyone else, except industry representatives.
- Industry – €850

LATE REGISTRATION – UNTIL JULY 1, 2023, AT 5 PM (CEST), OR EARLIER IF WE REACH CAPACITY

- Post graduate students – €180
  Young researchers who are still in training (students)
- Established researchers (including post docs) – €340
  Everyone else, except industry representatives.
- Industry – €1000

Please note that on-site registration will not be possible.

Registration includes:
- Participation at all ENCALS lectures and poster sessions
- Programme booklet
- Lunch on Wednesday, Tuesday and Friday
- Coffee/tea and something savoury during the breaks
- ENCALS gala dinner – sponsored by ENCALS

MAIN MEETING

The scientific meeting is at **CCIB - Centre de Convencions Internacional de Barcelona**

Maps, directions, a detailed accessibility guide and floorplans are available at the at the CCIB webpage: **https://ccib.es/es/**

WiFi: There is a special network for the meeting.
ENCALS Barcelona Meeting and password: **ENCALS2023**

If you have indicated that you have special dietary requirements, please make yourself known to a member of staff.

The registration desk will be staffed throughout the main meeting. Please also look out for our volunteers, who will be wearing yellow name badges. We are happy to help answer your questions.

There is a speakers' preparation room on **Floor 1, Room 118-119**. Here you can upload or check your presentation

There will be a cloakroom at the entrance of the venue where you can leave your belongings without any problem.

The dress code for the conference as a whole is casual, as is usual for ENCALS. Wear what makes you feel comfortable, but no onesies.

GALA DINNER

The Gala Dinner will be at the Recinto Modernista de Sant Pau on Thursday 13th July, at 20.15 h.

When you pick up your badge at the registration area, you will need to go to the Technical Secretariat, who will be right next to it, to confirm your attendance at the dinner.

You will be given a ticket which you must bring with you.
In case you need it, you can buy an extra ticket at the Technical Secretariat.

Tickets are limited, so please collect your ticket as soon as possible.

If you take a ticket, but later find yourself no longer able to attend, please return your ticket to the Technical Secretariat desk at the earliest opportunity, so that we can give it to someone on the waiting list.

The dress code for dinner is smart-casual.
You can get dressed up if you wish to, but this is not compulsory. Wear what you feel great in.

COMMITTEE MEMBERS

HOST
Mònica Povedano

LOCAL ORGANISING COMMITTEE


Mònica Povedano


Danique van der Gaauw

Torres Pardo Team

Maria Unzué

Carolina Rovira

Lidia Faucon

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SPONSORS

ENCALS would like to thank the following sponsors for their generous support of this year's meeting.

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PROGRAM AT A GLANCE

Monday July 10th

	ROOM 127-128	ROOM 131-132	ROOM 120-121	ROOM 129-130
08.00 - 09.00 h				
09.00 - 10.00 h	**PRECISION ALS** (09:30-19:00)			
10.00 - 11.00 h				
11.00 - 12.00 h				
12.00 - 13.00 h				
13.00 - 14.00 h				
14.00 - 15.00 h				
15.00 - 16.00 h				
16.00 - 17.00 h				
17.00 - 18.00 h				
18.00 - 19.00 h				

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Tuesday July 11th

PROGRAM AT A GLANCE

	ROOM 127-128	ROOM 131-132	ROOM 120-121	ROOM 129-130
08.00 - 09.00 h				
09.00 - 10.00 h	**INARC** (09:30-16:00)	**PRESYMPTOMATIC C9 COHORT STUDY** (10:00-14:30)	**EUPALS ANNUAL GENERAL MEETING** (09:00-12:00)	
10.00 - 11.00 h				
11.00 - 12.00 h				
12.00 - 13.00 h			**EUPALS BOARD OF DIRECTORS** (12:00-13:00)	
13.00 - 14.00 h				
14.00 - 15.00 h				**THIERRY LATRAN FOUNDATION** (14:00-19:30)
15.00 - 16.00 h		**DIGITAL HEALTHCARE TECHNOLOGY FOR ALS** (15:30-18:00)	**EUPALS ROUND TABLE** (16:00-19:00)	
16.00 - 17.00 h				
17.00 - 18.00 h				
18.00 - 19.00 h				

PROGRAM AT A GLANCE

Wednesday July 12th

	MAIN ROOM 111-112	ROOM 127-128	ROOM 131-132	ROOM 129-130
08.00 - 09.00 h		**ZAMBON BIOTECH** (08:00-09:30)	**WAVE LIFE SCIENCES** (08:00-09:30)	
09.00 - 10.00 h				**CORCEPT** (08:45-10:45)
10.00 - 11.00 h		**SANOFI** (10:30-12:00)	**BRAINTALE & ITALFARMACO GROUP** (10:30-12:00)	
11.00 - 12.00 h				**AMYLYX** (11:45-12:45)
12.00 - 13.00 h	**LUNCH** (12:00-12:45)			
13.00 - 14.00 h	**OPENING CEREMONY** (12:45-13:20)			
14.00 - 15.00 h	**SESSION 1. MOLECULAR MECHANISMS (PART I)** (13:20-14:40)			
15.00 - 16.00 h	**BREAK** (14:40-15:10) **SESSION 2. MOLECULAR MECHANISMS (PART II)** (15:10-16:20)			
16.00 - 17.00 h	**BREAK** (16:20-16:50)			
17.00 - 18.00 h	**SESSION 3. PROTEOMICS** (16:50-18:00)			
18.00 - 19.00 h	**POSTER SESSION 1 WITH WINE AND CHEESE** (18:00-19:30)			
19.00 - 20.00 h				

PROGRAM AT A GLANCE

Thursday July 13th

	MAIN ROOM 111-112	ROOM 127-128	ROOM 131-132	ROOM 129-130
08.00 - 09.00 h				
09.00 - 10.00 h	**SESSION 4. GENOMICS AND TRANSCRIPTOMICS** (08:30-09:40)			
	BREAK (09:40-10:10)			
10.00 - 11.00 h	**SESSION 5. THERAPEUTICS** (10:10-11:30)			
11.00 - 12.00 h	**BREAK** (11:30-11:50)			
12.00 - 13.00 h	**DEBATE. EARLY TO MARKET OR DEFINITIVE EVIDENCE** (11:50-12:35)			
13.00 - 14.00 h	**LUNCH** (12:35-13:45)			
14.00 - 15.00 h	**SESSION 6. NEUROPATHOLOGY AND APPLIED CLINICAL NEUROSCIENCE** (13:45-15:15)			
15.00 - 16.00 h	**BREAK** (15:15-15:45)			
	BASIC RESEARCH RAPID FIRE (1/2) (15:45-16:15)			
16.00 - 17.00 h	**SESSION 7. EPIDEMIOLOGY** (16:15-17:40)			
17.00 - 18.00 h	**BREAK** (17:40-17:50)			
18.00 - 19.00 h	**POSTER SESSION 2 WITH WINE AND CHEESE** (17:50-19:00)			

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Friday July 14th

PROGRAM AT A GLANCE

	MAIN ROOM 111-112	ROOM 127-128	ROOM 131-132	ROOM 129-130
08.00 - 09.00 h				
09.00 - 10.00 h	**SESSION 8. APPLIED NEUROPHYSIOLOGY** (08:45-10:00)			
10.00 - 11.00 h	**TRANSLATIONAL AND CLINICAL RAPID FIRE (2/2)** (10:00-10:40)			
	BREAK (10:40-11:10)			
11.00 - 12.00 h	**YOUNG INVESTIGATOR AWARDS** (11:10-11:55)			
12.00 - 13.00 h	**SESSION 9. PRE-SYMPTOMATIC PHENOTYPE AND COGNITION** (11:55-13:15)			
13.00 - 13.30 h	**CLOSING: NEXT ENCALS MEETING + PRIZES** (13:15-13:30)			

SCIENTIFIC PROGRAM

SCIENTIFIC PROGRAM | Wednesday 12-07-2023

10:00-12:00 h Registration
12:00-12:45 h Lunch

12:45-13:20 h Opening
Monica Povedano & Leonard van den Berg

13:20-14:40 h Session 1. Molecular mechanisms (Part I)
Ludo van den Bosch & Pascual Torres
Invited speaker: *Dr. Potero Otin*
Lipid Landschapes in ALS

1. The role of SPP1 and perivascular fibroblasts in ALS neurodegeneration
Jianing Lin, Karolinska Institutet (Sweden)
2. TDP-43 loss induces cryptic 3'end processing in neurons and ALS brains
Sam Bryce-Smith, UCL Queen Square Motor Neuron Disease Centre (United Kingdom)
3. Involvement of oligodendrocytes in Amyotrophic Lateral Sclerosis (ALS) and Frontotemporal Dementia (FTD) linked to Fused in Sarcoma protein (FUS)
Marguerite Jamet, University of Strasbourg (France)
4. Hexanucleotide repeat expansions in C9orf72 alter microglial responses and prevent a coordinated glial reaction in ALS
Philip Van Damme, University of Leuven (Belgium)

14:40-15:10 h Break

15:10-16:20 h Session 2. Molecular Mechanism (Part II)
Susanna Petri & Ruben Lopez Vales
Invited speaker: *Ana Martínez*
TDP43 cell-to-cell propagation in human sporadic ALS immortalized lymphocytes

5. NEK1 loss-of-function mutation impairs ciliogenesis in iPSC-motoneurons
Marta Nice Sorce, Istituto Auxologico Italiano (Italy)
6. CRISPR/Cas9 screen in human iPSC-derived cortical neurons identifies NEK6 as a novel disease modifier of C9orf72 poly(PR) toxicity
Wenting Guo, KU Leuven-Stem Cell Institute (SCIL) (Belgium)
7. SIRT1 upregulation mitigates DPR induced toxicity in C9orf72-associated disease models
Sophie Imhof, Medical University of Vienna (Austria)
8. TBK1 loss-of-function is associated with cell autonomous microglial dysfunction
Uroosa Chughtai, Cardiff University (United Kingdom)

16:20-16:50 h Break

16:50-18:00 h Session 3. Proteomics
Caroline Ingre & Alberto Ortega Cano
Invited speaker: *Janice Robertson*
The Hidden Mechanisms: Shedding Light on C9orf72 Loss of Function in ALS and FTD

9. Characterization of the human spinal cord synaptic proteome in ALS
Zsofia Laszlo, University of Dundee (United Kingdom)
10. Characterising the cortical synaptic proteome of Amyotrophic Lateral Sclerosis (ALS)
Nicole Hindley, University of Dundee (United Kingdom)
11. Multiomic profile integration reveals early disease signatures of Amyotrophic Lateral Sclerosis
Paul Lingor, Technical University of Munich (Germany)
12. Early mechanisms of neurotoxicity associated with intercellular spreading of TDP-43 pathology
Laura Rodríguez Gómez, Biodonostia Research Institute (Spain)

18:00-19:30 h Poster session 1 with wine and cheese

SCIENTIFIC PROGRAM | Thursday 13-07-2023

08:30-09:40 h **Session 4. Genomics and Transcriptomics**
Janine Kirby & Russell McLaughlin & Garcia Redondo
Invited speaker: *Abraham Acevedo*
New mouse models of TDP-43 proteinopathies

13. A Single-nuclei RNA Sequencing Approach of Oligodendrocyte Dysfunction in ALS
Maria Georgopoulou, Leuven Brain Institute (LBI) (Belgium)
14. Untangling the role of microRNAs in ALS pathogenesis via the iPSC-derived skeletal muscle in vitro model derived from C9ORF72-mutant patients
Claudia Malacarne, RCCS Istituto Neurologico Carlo Besta (Italy)
15. Sex-stratified analysis of ~133k samples identifies novel associations with Amyotrophic Lateral Sclerosis
Ross Byrne, Trinity College Dublin (Ireland)
16. Genome-wide methylation array reveals epigenetic drift and epivariations in ALS
Alberto Brusati, University of Pavia (Italy).

09:40-10:10 h Break

10:10-11:20 h **Session 5. Therapeutics**
Thomas Meyer & Angela Genge
Invited speaker: *Neil Shneider*
New therapeutic landschape

17. Assessment of the therapeutic effect of IGS 2.7, a CK1 protein kinase inhibitor, in combination with riluzol for the treatment of ALS
Loreto Martinez-Gonzalez, CSIC (Spain)
18. The MIROCALS Study: Efficacy of low dose IL2 in ALS and implications for ALS trial design
Nigel Leigh, Brighton and Sussex Medical School (United Kingdom)
19. Lys-acetylated PPIA as a candidate therapeutic target for TDP-43 proteinopathies
Laura Pasetto, Istituto di Ricerche Farmacologiche Mario Negri IRCCS, (Italy)
20. Neurofilament light chain response during therapy with Tofersen in SOD1-related ALS – treatment experience in clinical practice
Thomas Meyer, Universitätsmedizin Berlin (Germany)
21. Evaluating single and multiple ascending doses of WVE-004 in C9orf72-associated ALS and FTD: results from the FOCUS-C9 Trial
Michael Tillinger, Wave Life Sciences (USA)

11:30-11:50 h Break

11:50-12:35 h **Debate. Early to market or Definitive evidence**
Leonard van den Berg
Speakers: *Jinsy Andrews and Chris McDermott*

12:30-13:45 h Lunch

13:45-15:15 h **Session 6. Neuropathology and applied clinical neuroscience**
Ammar Al-Chalabi & Caco Vazquez Costa
Invited speaker: *Andrea Malaspina*
Emerging biomarkers and evidence from ALS clinical trials

22. Metabolic Alterations precede Neurofilament Changes in Presymptomatic ALS Gene Carriers
Johannes Dorst, University of Ulm (Germany)
23. Disruption of the Angiopoietin-like protein system correlates with lipid homeostasis in ALS
Sruthi Sankari Krishnamurthy, University of Ulm (Germany)
24. High-resolution assessment of ALS neuropathology and its association with clinical presentation
Anna Sanchez Avila, University of Dunde (United Kingdom)

SCIENTIFIC PROGRAM | Thursday 13-07-2023

25. Investigating the role of autoantibodies against neurofilaments in amyotrophic lateral sclerosis (ALS)
Ellie Sturmey, Queen Mary University of London (United Kingdom)
26. Lipid-mediated resolution of inflammation in amyotrophic lateral sclerosis informs on novel biomarkers and therapeutic targets
Ozlem Yildiz, Queen Mary University of London (United Kingdom)
27. Unsupervised machine learning identifies distinct molecular and phenotypic ALS subtypes in post-mortem motor cortex and blood expression data
Heather Marriott, King's College London (United Kingdom)

15:15-15:45 h Break

15:45-16:15 h **Basic research Rapid Fire (1/2)**
Philip van Damme & Miguel Angel Rubio

i) Deletion of endothelial TDP-43 disrupts the vascular barrier triggering inflammation and hemorrhages in the central nervous system
Victor Arribas, University of Barcelona and Bellvitge Biomedical Research Institute (Spain)
ii) Haploinsufficiency of C9ORF72 selectively impairs autophagy in C9ORF72-linked ALS
Rim Diab, Inselspital University Hospital (Switzerland)
iii) Elucidating the timing of TDP-43 related phenotypes using iPSC-derived motor neurons from TARDBP ALS patients
Melissa Nijs, University of Leuven (Belgium)
iv) Dynamic Translational Profile of Stressed iPSC-MNs from C9orf72-ALS Patients by Translating Ribosome Affinity Purification (TRAP)
Yinyan Xu, University of Oxford (United Kingdom)
v) MAM lipidome changes associated with TDP-43 dysfunction
Anna Fernàndez Bernal, University of Lleida- IRB Lleida (Spain)
vi) Gap junctions are functionally enhanced in iAstrocytes derived from C9ORF72 repeat expansion *patients*
Iris Stefania Pasniceanu, University of Sheffield (United Kingdom)
vii) Targeting De Novo Fatty Acid Synthesis as a Therapeutic Strategy to Alleviate Non-cell Autonomous Mechanisms in ALS
Maddi Garciandia, Biodonostia Health Research Institute (Spain)
viii) Epigenetic analysis on organoids for the study of Amyotrophic Lateral Sclerosis
Eveljn Scarian, IRCCS Mondino Foundation (Italy)
ix) Cognate microglia – T cell interaction induce neurotoxic T cell function in a fast-progressing C9orf72 ALS animal model
Qihui Zhou, German Center for Neurodegenerative Diseases (Germany)
x) An interaction between synapsin and C9orf72 regulates excitatory synapses and is impaired in ALS/FTD
Claudia Bauer, University of Sheffield, (United Kingdom)
xi) Cortical network dysfunction in ALS using task-free magnetoencephalography
Michael Trubshaw, University of Oxford (United Kingdom)

16:15-17:40 h **Session 7. Epidemiology**
Orla Hardiman & Markus Weber
Invited speaker: *Giancarlo Logroscino*
LAENALS new and old questions on ALS risk in diverse populations in Latin America

28. Genetic and Environmental Risk Factors contribute in the pathogenesis of ALS in Cyprus
Ellie Mitsi, The Cyprus Institute of Neurology and Genetics (Cyprus)
29. Incidence, mortality and survival of patients with amyotrophic lateral sclerosis (ALS) in Belgrade, Serbia (1994-2018)
Aleksa Palibrk, University Clinical Center of Serbia (Serbia)

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30. Presymptomatic geographical distribution of ALS patients suggests the involvement of environmental factors in the disease pathogenesis
Rosario Vasta, University of Turin (Italy)
31. Epidemiological Trends of Amyotrophic Lateral Sclerosis (ALS) in Ireland 1996-2021
Robert McFarlane, Trinity College Dublin (Ireland)
32. Population-level penetrance of ALS genes is markedly reduced
Andrew Douglas, Oxford University Hospitals NHS Foundation Trust (United Kingdom)
33. Epidemiology studies in Colombia
Martha Pena

17:40-17:50 h Break

17:50-19:00 h Poster session 2 with wine and cheese

20:15 h ENCALS Dinner Party

SCIENTIFIC PROGRAM | Friday 14-07-2023

08:45-10:00 h **Session 8. Applied Neurophysiology**
Roisin McMackin & Adolfo Lopez de Munain
Invited speaker: *Xavi Navarro*
Electrophysiological tests as biomarkers for ALS experimental research

34. Acceptability and feasibility of the MiNDToolkit intervention for management of behavioural symptoms in MND: the views of healthcare professionals
Thando Katangwe-Chigamba, University of East Anglia (United Kingdom)
35. M50, CMAP50 and MUNIX200 are potentially new parameters to describe disease progression in Amyotrophic Lateral Sclerosis
Annekathrin Roediger, Jena University Hospital (Germany)
36. Corticomuscular coherence in ALS during the performance of a motor task
Saroj Bista, Trinity College Dublin (Ireland)
37. Classification of ALS Patients Based on Resting-state EEG Trajectories: Clinical Relevance and Network Progression"
Marjorie Metzger, University of Dublin (Ireland)

10:00-10:40 h **Translational and clinical rapid fire (2/2)**
Adriano Chio & Ana Calvo

xii) isomiRs - a novel family of molecular biomarkers for ALS prognostication
Yahel Cohen, Weizmann Institute of Science (Israel)
xiii) Performance of serum neurofilament light chain in a wide spectrum of clinical courses of ALS – a cross-sectional multicenter study
Thomas Meyer, Universitätsmedizin Berlin (Germany)
xiv) Ultrasound-mediated blood–spinal cord barrier opening prolongs survival in an ALS mouse model
Ilyes Aliouat, Sorbonne Université (France)
xv) Motor system connectivity in ALS: A corticomuscular magnetoencephalography study
Katie Yoganathan, University of Oxford (United Kigndom)
xvi) Dysfunction Of Cortical Inhibitory Interneurons In Amyotrophic Lateral Sclerosis
Cristina Benetton, Sorbonne Université (France)
xvii) Nerve excitability disentangled: hyperexcitability in ALS is driven by altered slow potassium channel kinetics
Diederik Stikvoort Garcia, University Medical Center Utrecht (The Netherlands)
xviii) Social Cognition Impairment in Amyotrophic Lateral Sclerosis
Sara Kadenšek, University Medical Centre Ljubljana (Slovenia)
xix) Motor band sign is a specific marker of ALS and corresponds topographically to motor symptoms
Charlotte Zejlon, Karolinska University Hospital (Sweden)
xx) Investigating cognitive endophenotypes and presymptomatic cognition in unaffected relatives of familial ALS patients: a longitudinal study
Colm Peelo, Trinity College Dublin (Ireland)
xxi) Cortical network dysfunction in ALS using task-free magnetoencephalography
Michael Trubshaw, University of Oxford (United Kingdom)

10:40-11:10 h Break

11:10-11:55 h **Young Investigator Awards**
Ammar Al-Chalabi

SCIENTIFIC PROGRAM | Friday 14-07-2023

11:55-13:15 h **Session 9. Pre-symptomatic phenotype and cognition**
Raquel Sánchez del Valle & Ratko Radakovic
Invited speaker: *Caroline McHutchinson*
The temporal course of cognitive and behavioural changes in Motor Neuron Diseases

38. Phenoconversion of asymptomatic genetic mutation carriers to ALS/FTD: a longitudinal neuroimaging study
Kevin van Veenhuijzen, University Medical Center Utrecht (The Netherlands)
39. Investigation of cognitive networks in asymptomatic C9orf72 repeat expansion carriers using high-density EEG
Stefan Dukic, University Medical Center Utrecht (The Netherlands)
40. Cognitive impairment and capacity to consent to clinical trials in ALS
Debbie Gray, University of Edinburgh (United Kingdom)
41. Premorbid brain structural variation influences risk of ALS
Alexander Thompson, University of Oxford (United Kingdom)
42. Brain "neurovascular coupling" in amyotrophic lateral sclerosis: the link with cognitive impairment
Minoo Sharbafshaaer, Università degli Studi della Campania Luigi Vanvitelli (Italy)

13:15-13:30 h Closing: next ENCALS meeting + prizes

13:30 h Lunch

SYMPOSIA

ZAMBON BIOTECH
Wednesday July 12th | 08:00-09:30 h
Room 127-128
Tailoring oral disease-modifying treatment in Amyotrophic Lateral Sclerosis (ALS): a patient-centric, personalised approach

Introduction
Adriano Chiò, Italy
Oral disease-modifying therapy of ALS: what is standard of care?
Juan Francisco Vázquez-Costa, Spain
The challenges of oral disease-modifying therapy due to disease progression
Susanne Petri, Germany
What patients really need regarding oral disease-modifying therapy
Vincenzo Silani, Italy
Panel Discussion and Q&A

AMYLYX
Wednesday July 12th | 11:45-12:45 h
Room 129-130
Navigating the complexities of ALS clinical trials: clinical endpoints and clinical relevance
Chair: Juan Francisco Vázquez Costa

Speaker 1: Ruben van Eijk

Speaker 2 Adriano Chio

BRAINTALE & ITALFARMACO GROUP
Wednesday July 12th | 10:30-12:00 h
Room 131-132
Race against time : How to foster ALS diagnosis and holistic care ?
Chair: Pierre-François Pradat, France

The role of Neurophysiology in diagnosis and disease progression
Markus Weber, Switzerland
Unlocking Neuroimaging as an ALS biomarker
Pierre-François Pradat, France
Why is it important to start the treatments at an early stage of the disease in ALS ?
Christian Lunetta, Italy
Keeping patients moving - The patient perspective on physical therapy and motor assisted movement exercisers
Andre Maïer, Germany

CORCEPT
Wednesday July 12th | 08:45-10:45 h
Room 129-130
DAZALS Meeting

By Invitation Only (for DAZALS Investigators and Site Staff)

SANOFI
Wednesday July 12th | 10:30-12:00 h
Room 127-128
Uncovering RIPK1 as a Potential Target in ALS

Understanding the Unmet Needs in ALS
Professor Leonard van den Berg
Introduction to RIPK1 Biology
Professor Jeffrey Rothstein
The Role of RIPK1 in ALS Pathophysiology
Professor Junying Yuan
Summary
Professor Jeffrey Rothstein and Professor Leonard van den Berg
Q&A Session
Moderated by Professor Jeffrey Rothstein and Professor Leonard van den Berg
Questions presented to all faculty
Close
Professor Leonard van den Berg

WAVE LIFE SCIENCES
Wednesday July 12th | 08:00-09:30 h
Room 131-132

Wave Life Sciences Investigator Meeting, by invitation only

TOGETHER for ALS

ALS: a disorder with a devastating physical and psychological impact

People living with ALS face rapid and relentless loss of physical functions, including deteriorating muscle function, loss of ability to move, speak and breathe unaided and eventually, death. This loss of independence has a devastating emotional impact, not only on those living with the disorder but also their loved ones. Its impact, however, is not well known by the public.

#TogetherForALS aims to amplify the voices of people living with ALS, their loved ones and the community by sharing their stories.

Support the campaign & get involved

#1 Visit the website and watch the videos

#2 If you are a patient advocacy group and would like to get involved, contact us via email using info@amylyx.com

Scan the QR code to visit www.TogetherForALS.com and hear stories from the ALS community

ALS, amyotrophic lateral sclerosis

AMYLYX

We are in this together.

#TogetherForALS

We're working toward more moments like this

In memory of Mick, a husband and father, who was a gifted tattoo artist and musician.

AMYLYX

NP-03523 | May 2023

Biogen is proud to sponsor the 2023 ENCALS meeting.

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May 2023. Biogen-211234.

At Biogen, we are pioneering new science that takes us deep into the body's nervous system, and stretches wide across digital networks and patient communities, to better understand, and preserve, the underlying qualities of our essential human nature.

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May 2023. Biogen-211225.

Committed to supporting ALS patients through clinical research.

[redacted] founded by [redacted], is a private company founded in [redacted] dedicated to the manufacture and marketing of quality pharmaceutical products.

We are committed to the development of medicines to improve patients' lives and quality of life in [redacted], neurology, diabetes, gastroenterology and new diseases.

[redacted] is currently active in development for the treatment of patients with Amyotrophic Lateral Sclerosis.

BRUSCHETTINI

Cytoskeleton

Contractions aren't just for grammar.


Every day we are motivated by people living with diseases of impaired muscle function. They are not defined by their disease. They are fighting with spirit, determination and courage. They amaze us. They inspire us. They are our heroes.
Patients are our North Star

For PTC Therapeutics, our 25-year history has centered on bold, transformative science that brings more moments to patients and their families living with rare diseases.

TRANSLATING SCIENCE

Transforming Lives

Extending life's moments for people living with rare diseases

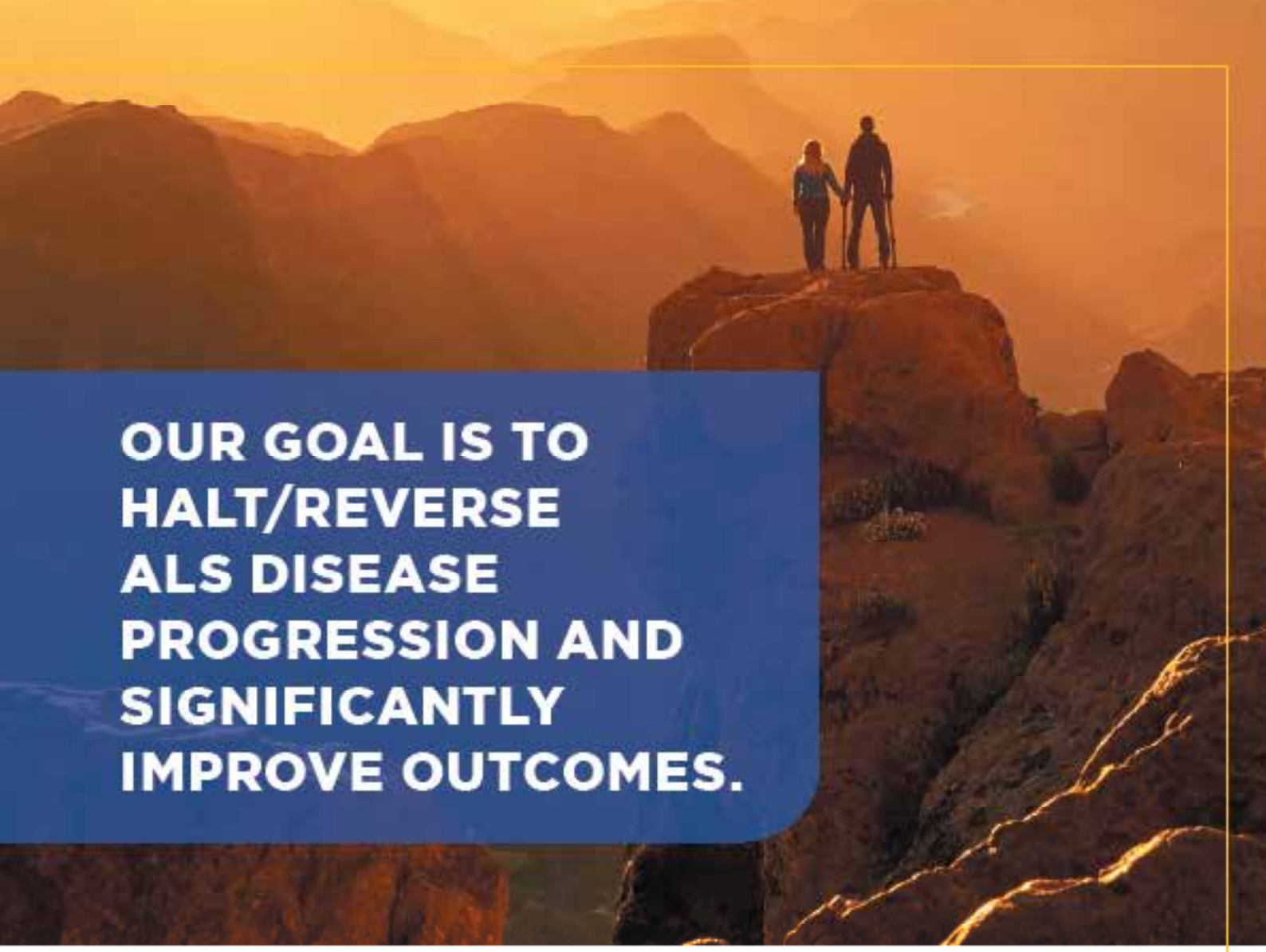
PTC 25 THE EXPERIENCE

No matter how uncommon the disorder, the life-limiting effects are a daily reality for those affected.

That's why we're researching life-changing treatments every day, including an investigational treatment for amyotrophic lateral sclerosis (ALS).

Learn more at clinicaltrials.ptcbio.com.

PTC
THERAPEUTICS



QurAlis is a proud sponsor of the ENCALS Meeting 2023

At QurAlis, we are pioneering the path to conquering ALS and other neurodegenerative diseases with genetically validated targets with next-generation precision medicines. Our proprietary platforms and unique biomarkers enable the design and development of precision medicines that act directly on disease-causing genetic alterations.

Our mission is to make a meaningful difference in patients' lives.

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QurAlis

QurAlis.com

Light The Way

Powered by Sano

These at risk of genetic ALS/MND often struggle to receive the education, support, and access to genetic testing they need to make informed decisions.

We see you, and we want to help.

If you're an industry partner (e.g. sponsor, physician or patient advocacy group) interested in finding out more please contact us below:

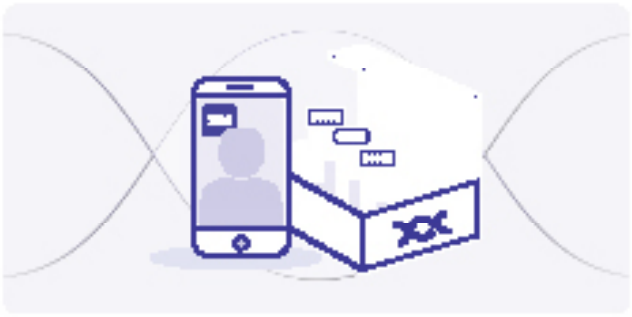
https://sanogenetics.typeform.com/light-the-way

Light the Way is a free online platform, supported by grant funding and industry partners, launching Autumn 2023 that will offer:

- ATXN2
- TARDBP
- C9orf72
- FUS
- SOD1

Access to free, comprehensive genetic testing, screening over 20 genes including SOD1, C9orf72, FUS, ATXN2, TARDBP

Access to free genetic counselling from a trained professional, both before and after testing



The opportunity to discover relevant clinical trials

Access to expertly developed educational materials

- BEACON by Sano Genetics

Participation in Beacon, a survey-based longitudinal research study which aims to inform future policy. The study will monitor mental well-being following the receipt of genetic test results.

Launching autumn 2023 in the UK and USA (in English and Spanish)

For industry partners to learn more about our multi-sponsor patient-finding program and registry:

Submit a get in touch form to speak with our team
https://sanogenetics.typeform.com/light-the-way

At the event, speak with Dr. Paul White
paul.white@sanogenetics.com

Delivering on the promise of gene therapy

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Innovating to transform the lives of patients

uniQure.com

Uncovering RIPK1 as a Potential Target in ALS

Wednesday, 12 July 2023, 10:30–12:00 CEST
ENCALS 2023, Centre de Convencions Internacional de Barcelona, Room 127–128, Barcelona, Spain

Expert Speakers

Professor Leonard van den Berg, MD, PhD (Chair)

Neurology and Neuroscience, UMC Utrecht, Utrecht, Netherlands

Professor Jeffrey Rothstein, MD, PhD (Co-Chair)

Neurology and Neuroscience, Johns Hopkins School of Medicine, Maryland, USA

Professor Junying Yuan, PhD

Interdisciplinary Research Center on Biology and Chemistry, Shanghai Institute of Organic Chemistry, Shanghai, China

Agenda

Time (CEST)	Duration	Topics	Presenter
10:30–10:45	15 minutes	***Understanding the Unmet Needs in ALS***	Professor Leonard van den Berg
10:45–10:55	10 minutes	***Introduction to RIPK1 Biology***	Professor Jeffrey Rothstein
10:55–11:25	30 minutes	***The Role of RIPK1 in ALS Pathophysiology***	Professor Junying Yuan
11:25–11:30	5 minutes	***Summary***	Professor Jeffrey Rothstein and Professor Leonard van den Berg
11:30–12:00	30 minutes	***Q&A Session***	Moderated by Professor Jeffrey Rothstein and Professor Leonard van den Berg. Questions presented to all faculty
12:00		***Close***	Professor Leonard van den Berg

This meeting is organised and sponsored by

sanofi

Dear colleagues,

We are pleased to invite you to join our expert faculty for Sanofi's first-ever ALS symposium titled ***'Uncovering RIPK1 as a Potential Target in ALS'*** *at ENCALS 2023 in Barcelona for an in-person event on* ***Wednesday 12 July 2023 at 10:30–12:00 CEST.***

Together with our esteemed colleague ***Professor Junying Yuan****, Interdisciplinary Research Center on Biology and Chemistry, Shanghai Institute of Organic Chemistry (Shanghai, China),* ***we will discuss the role of RIPK1 in neuronal inflammation and cell death pathways, including its potential link to ALS pathophysiology****. We will share our experiences and knowledge with you and encourage your participation to facilitate an engaging and educational session.*

We look forward to your participation in our symposium and cannot wait to welcome you!

Professor Leonard van den Berg, MD, PhD (Chair)

Neurology and Neuroscience,
UMC Utrecht, Utrecht, Netherlands

Leonard van den Berg

Professor Jeffrey Rothstein, MD, PhD (Co-Chair)

Neurology and Neuroscience,
Johns Hopkins School of Medicine, Maryland, USA

jeffrey rothstein

sanofi

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Zambon invites you to its satellite symposium

8:00 am on the **12th July** in **Room 127-128** (floor P1)

- *Adriano Chiò: Full Professor of Neurology; Chair, Scientific Board, Ferdinando Rossi School of Advanced Studies of Turin; Director, Neurology 1 Division, Città della Salute e della Scienza Hospital, Turin; Director, ALS Regional Expert Center of Turin Rita Levi Montalcini, Department of Neuroscience, University of Turin, Italy*
- *Susanne Petri: Associate Professor of Neurology; Interim Director, Department of Neurology, Hannover Medical School, Germany*
- *Vincenzo Silani: Former Full Professor of Neurology – University of Milan; "Dino Ferrari" Center – University of Milan; Director Department of Neuroscience and Director Experimental Laboratory of Neuroscience – IRCCS Istituto Auxologico Italiano, Milan, Italy*
- *Juan Francisco Vázquez Costa: Coordinator ALS Unit, Hospital la Fe, Valencia, Spain; Associate Professor of Neurology, Universitat de Valencia; Member of CIBERER (Spanish research network for rare diseases), Spain*

During this 90-minute session, experts in the field of amyotrophic lateral sclerosis will discuss approaches to oral disease-modifying treatment, revealing some of the barriers that prevent patients from experiencing the treatment continuity they require to optimally manage their disease – together with some potential solutions.
We hope you will join us for this series of educational and thought-provoking presentations.

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Quick access after the sessions through ENCALS Newsletters, don't miss out!

Rare but still there

Recognise the dragon. Recognise ALS.

Having Amyotrophic Lateral Sclerosis (ALS) can feel like your energy is slowly burning out.

Unfortunately, the condition can be overlooked, which significantly delays diagnosis and causes serious consequences.

The phantasticALS campaign was created to make all the dragons fighting ALS more seen and to encourage all to join us in raising awareness about this rare and serious condition.

Join us in reminding everyone that while people with ALS might be rare, they're still there.

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ENCALS

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DISCOVER OUR FNP-122 CLINICAL DEVELOPMENT PROGRAM

PIVOTAL PHASE 3

ADORE (ALS trial with Daily ORal Edaravone)

A multicenter, randomized, double-blind, placebo-controlled study to investigate the efficacy and safety of FNP122 in patients with Amyotrophic Lateral Sclerosis (pALS).

300 pALS

FNP122 (daily oral edaravone) vs. Placebo - 2:1 ratio to reduce placebo burden

- Multicenter
- Randomized
- Multinational
- Placebo-controlled
- Double-blind

Main Inclusion criteria

- Definite, probable, probable laboratory supported, or possible ALS (El Escorial and the revised Airlie House diagnostic criteria)
- Onset of first symptoms ≤24 months prior to randomization
- Change ≥0.35 and ≤1.5 points per month in ALSFRS-R score between first symptoms and screening visit
- Slow Vital Capacity (SVC) ≥70%

Endpoints

Primary efficacy endpoint

Secondary efficacy endpoints

Safety endpoints

Exploratory endpoints

Study design

300 patients

2:1

200 patients - daily oral edaravone (FNP122) + Riluzole

100 patients - matched placebo + Riluzole

Conclusions

The ADORE trial is an ongoing phase 3 study designed to compare active treatment vs. placebo for a longer duration (up to 72w) in pALS to provide relevant safety and efficacy data on this new daily oral formulation of edaravone (FNP122).

Neuroscience Hub

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https://www.ferrer.com/en/adore-graphical-abstract

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TO BRING DAILY ORAL EDARAVONE TO EUROPE

OPEN LABEL EXTENSION

ADORE (ALS trial with Daily ORal Edaravone EXTension)

A multicenter, open-label extension study to investigate the long-term safety of FNP122 in patients with Amyotrophic Lateral Sclerosis.

TRICALS

European Study conducted with TRICALS support

Inclusion criteria

Endpoints

Primary endpoint

Secondary endpoints

Safety endpoints

Study design

Baseline	Visits every 2 months	End of trial

Conclusions

The ADOREXT study will provide relevant safety and long-term survival data on this daily oral formulation of edaravone (FNP-122)

Neuroscience Newsletter

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Poster titles and authors

Basic research

1. 3D in vitro brain and spinal cord cell models to identify early neuronal vulnerability and test therapies in C9ORF72-Amyotrophic Lateral Sclerosis

Delia Gagliardi(1)*, Andrea D'Angelo(1), Francesca Beatrice(2), Paola Rinchetti(1), Chiara Cordiglieri(3), Giacomo Pietro Comi(1,4), Monica Nizzardo(2), Linda Ottoboni(1), Stefania Corti(1,2)

2. A Novel Gene Therapy Approach for ALS by Overexpressing the Pleiotropic Chronokine α-Klotho

Sergi Verdés (*)(1)(2)(5), Mireia Herrando-Grabulosa (1)(3)(4), Rubén Guerrero-Yagüe (1)(2), Joan Roig-Soriano (1)(2)(5), Núria Gaja-Capdevila (1)(3)(4), Judith Sauleda (1)(2), Marc Leal-Julià (1)(2)(5), Andrea Onieva (1)(2)(5), Laura Rodriguez-Estevez (1)(2)(5), Javier del Rey (1)(2), Neus Hernández (1)(3), Miguel Chillón (1)(2)(5)(6), Xavier Navarro (1)(3)(4), Assumpció Bosch (1)(2)(4)(5)

3. A repurposing approach to improve disease outcome in SOD1G93A mice by counteracting oligodendrocyte dysfunction and neuroinflammation

Stefano Raffaele (1), Nhung Nguyen (2), Marta Boccazzi (3), Giulia Frumento (2), Marco Milanese (2,4), Francesca C. Mannella (1), Kate L. Lambertsen (5,6,7), Kirsten Madsen (8), Henrik D. Schroeder (8), Eva K. Hejbøl (8), Giambattista Bonanno (2,4), Maria P. Abbracchio (3), Tiziana Bonifacino (2,4), Marta Fumagalli (1)*

4. Activation of Boron transporter (NaBC1) in muscle generates neuroprotection in a SOD1 ALS mouse model.

Ana Rodríguez-Romano (1), Jorge Vinagre (1*), Patricia Muñoz (1), Laura Moreno-Martinez (2), Juan Francisco Vázquez Costa (3), Rosario Osta (2), Patricia Rico (1).

5. ALS is associated to disorders of riboregulation of p62 by vtRNAs

Santiago Rico-Rios*(1), Miriam Ceron-Codorniu (1), Pascual Torres (1), Victoria Ayala (1), Pol Andrés-Benito (2), Mònica Povedano (3), Isidro Ferrer (3), Reinald Pamplona (1), Manuel Portero-Otin (1)

6. ALS/FTD-associated C9orf72 C4G2 repeat RNA disrupts phenylalanine tRNA aminoacylation and protein production

Urša Čerček (1,2*), Mirjana Malnar (1,2), Xiaoke Yin ((3), Manh Tin Ho (4), Barbka Repic Lampret (5), Manuela Neumann (6,7), Andreas Hermann (8,9), Guy Rouleau (10,11), Beat Suter (4), Manuel Mayr (3), BorisRogelj (1,12)

7. ALS-associated GLT8D1 mutations disrupt membrane lipid rafts and localised neurotrophin signalling

Tobias Moll* (1,2), Emily Graves (1,2), Agnieszka Urbanek (1,2), Nikita Soni (1,2), Ramya Ranganathan (1,2), Adrian Higginbottom (1,2), Shanshan Wang (3), Andrew J Grierson (1,2), Brian P Head (3, 4), Johnathan Cooper-Knock (1, 2) & Pamela J Shaw (1,2)

8. Altered dynamics of DNA damage repair associated proteins in a human, neuronal in-vitro cell model of FUS-ALS

Marcel Naumann* (1), Anne Richter (2), Stephan W. Grill (3), Andreas Hermann(1,4,5)

9. An electrophysiological study on C9orf72 associated motor neuron disease

Omar Keritam1,2,*, Sophie Imhof1,2, Hakan Cetin1,2

10. An homozygous C9orf72 transgenic mouse model displays enhanced molecular hallmarks of ALS and reveals AS gene therapy ability to target these traits.

Georges Arielle Peche*(1), Marisa Cappella(1), Alessandra Ricupero(1), Benoit Giroux(1), Mathilde Cohen-Tannoudji(1), Stéphanie Astord(1), Sonia Pezet(1), Udit Sheth(2), Tania Gendron(2), Piera Smeriglio(1), Maria Grazia Biferi(1)

11. An in vitro investigation of cortical network hyperexcitability in mouse models of ALS

Marilina Douloudi*(1), Agnieshka Urbanek(1), Andrew Grierson(1), Jodie Watkins(2), Matthew Livesey(1), Richard Mead(1).

12. Analysis of stress response in Amyotrophic Lateral Sclerosis (ALS) patient fibroblasts

Sibylle de Bertier*(#)(1), François Muratet (#)(1), Maria-Del-Mar Amador (1,2), Elisa Teyssou (1), David Akbar (1), Beata Gyorgy (1), Justine Guégan (1), Yannick Marie (1), Ludmila Jornea (1), Stéphanie Bigou (1), Valentine Marquet (3), Sylvie Bourthoumieu (3), Catherine Yardin (3,4), Géraldine Lautrette (5), Philippe Couratier (5), Danielle Seilhean (1,6), Christian S Lobsiger (1), Séverine Boillée (1), Nadine Le Forestier (1,2,7), François Salachas (1,2), Delphine Bohl (1), Stéphanie Millecamps (1).

13. Antibody based treatment: Amelioration of motor and cognitive performances and reduction of TDP-43 proteinopathy in mice models of ALS induced by the

Amélie Poulin-Brière* (1), Jean-Pierre Julien (2), Silvia Pozzi (3)

14. Assessment of protein aggregation in a model of ALS and cells derived from patients

Carmen Pérez de la Lastra (1,2), Gracia Porras Franco (1,3), Carlota Tosat-Bitrián (1,3), Ángeles Martín-Requero (1,3), Ana Martínez Gil (1,3) and Valle Palomo (2,3)

15. Assessment of the therapeutic effect of IGS 2.7, a CK1 protein kinase inhibitor, in combination with riluzol for the treatment of ALS

Loreto Martinez-Gonzalez* (1,2), Carmen P. de la Lastra-Aranda (1), Marta Gomez-Almeria (2,3,4), Eva de Lago (2,3,4), Valle Palomo (2,5), Ana Martinez (1,2)

16. Avoiding TDP-43 cell-to-cell propagation in human sporadic ALS immortalized lymphocytes by treatment with IGS2.7, a CK-1 inhibitor

Ana Martinez* (1,2), Eva Pérez Cuevas (1), Loreto Martinez-Gonzalez (1,2), Carlota Tossat (1), Carmen Gil (1), Valle Palomo (2,3), Angeles Martin-Requero (1).

17. Axonal mRNA homeostasis in ALS – a link between DNA damage and mitochondria?

Hannes Glaß*(1), Vitaly Zimyanin (2,3), Banaja Dash (1), Julia Pfeiffer (1), Jette Abel (1), Andreas Hermann (1,4,5)

18. Beneficial effects of CB2 receptor activation at central and peripheral sites in ALS murine models

*Marta Gómez-Almería(1-3), Claudia Gonzalo-Consuegra (1-3), Carmen Rodríguez-Cueto (1-3), Matthias Wittwer (4), Pawel Dzygiel (4), Uwe Grether (4), Javier Fernández-Ruiz (1-3) and Eva de Lago (1-3)

19. Beta-adrenergic modulation of motoneuronal signaling, electrophysiology and disease-associated transcriptome in the SOD1-ALS mouse model

*Guillaume Caron (1) , Stefano Antonucci (1,2) , Natalie Dikwella (2), Marcin Bączyk (3), Albert Ludolph (2,4) Daniel Zytnicki (1) , Francesco Roselli (2,4)

20. Boosting the peripheral immune response in the skeletal muscles improved motor function in Amyotrophic Lateral Sclerosis (ALS) transgenic mice

Maria Chiara Trolese* (1), Carlotta Scarpa (1), Valentina Melfi (1), Paola Fabbrizio (1), Francesca Sironi (1), Martina Rossi (1), Caterina Bendotti (1), Giovanni Nardo (1).

21. C9ORF72 deficiency results in degeneration of the vertebrate retina in vivo

Andrea Salzinger (1,2), Natalia Jaroszynska (3), Themistoklis M. Tsarouchas (1), Catherina G. Becker (1), Thomas Becker (1), David A. Lyons (1), Ryan B. MacDonald(3), Marcus Keatinge*(1,2)

22. C9orf72-ALS iPSC microglia are pro-inflammatory and induce apoptosis of co-cultured wild-type motor neurons via MMP9

Björn F. Vahsen (*) (1,2), Sumedha Nalluru (1), Lucy Farrimond (1,2), Georgia Morgan (1), Benazir Amein (1), Emily Carroll (1,2), Yinyan Xu (1,2), Jakub Scaber (1,2), Mireia Carcolé (3), Adrian M. Isaacs (3), Elizabeth Gray (1), Martin R. Turner (1), Sally A. Cowley (4), Kevin Talbot (1,2)

23. Cellular and axonal transport phenotypes due to the C9orf72 HRE in iPSC-motor and sensory neurons

Jakub Scaber*(1)(2), Iona Thomas-Wright(1)(2), Alex Clark(1), Yinyan Xu(1)(2), Björn Vahsen(1)(2), Mireia Carcolé(3), Ruxandra Dafinca(1)(2), Lucy Farrimond(1)(2), Adrian Isaacs(3), David Bennett(1), Sally A. Cowley(4), Kevin Talbot(1)(2)

24. Cellular Stress induces Neuron-specific Degradation of the Fragile X Protein Family

Lorena Decker*(1), Sarah Müller(1), Sonja Menge(1), Albert C. Ludolph(1,2), Axel Freischmidt(1)

25. Characterising novel, humanised and physiological mouse models of FUS-ALS

Georgia Price*(1,2), Samanta Gasco(1,3), Remya R Nair(1,4), Charlotte Tibbit(1), Zeinab Ali(1), Dave Thompson(1,2), Anny Devoy(1,5), Marc David-Ruepp(5), Rosie Bunton-Stasyshyn(1), Gemma F Codner(1), Lydia Teboul(1), Alasdair Allan(1), Adele Austin(1), Jackie Harrison(1), Gemma Atkins(1), Elizabeth MC Fisher(1,6), Thomas J Cunningham(1,7)

26. Characterising pathological oligomers with single-molecule microscopy

Rebecca Saleeb*(1), Mathew Horrocks(1)

27. Characterization of a therapeutic approach to target TDP-43 proteinopathy using phage display-derived scfv

Yara Al Ojaimi (1)*, Rudolf Hergesheimer (1), Anna Chami (1), Hugo Alarcan (1,3), Johanna Augros (1), Stéphanie David (2), Emilie Allard-Vannier (2), Patrick Vourc'h (1,3), Christian Andres (1,3), Philippe Corcia (1,4), Astrid Musnier (5), Anne Poupon (5), Eric Reiter (5), Martine Pugnière (6), Olivier Herault (7), Pierre Martineau (6), Débora Lanznaster (1), Hélène Blasco (1,3)

28. Characterization of TDP-43 role in cellular models exposed to ALS-related stress

Miriam Ceron-Codorniu* (1), Pascual Torres (1), Santiago Rico-Rios (1), Anna Fernandez-Bernal (1), José CE Serrano (1), Jordi Boada (1), Reinald Pamplona (1), and Manuel Portero-Otín (1)

29. Characterization of the KIF5A aggregates in patient-derived induced pluripotent stem cells

(1,2)(*)Isabel Loss, (1)Rüstem Yilmaz, (1)Rosanna Parlato, (3)Johannes Wilbertz, (3)Loic Cousin, (3)Karen Schmitt, (2) Philipp Koch, (1)Jochen Weishaupt

30. CLIP-Seq Analysis Enables the Design of Ribosomal RNA Bait Oligonucleotides That Protect Against C9ORF72 ALS/FTD-Associated Poly-GR Pathophysiology

Juan A. Ortega(1, 2), Ivan R. Sasselli(3,4,5), Marco Boccitto(6), Andrew C. Fleming(1), Tyler R. Fortuna(7), Yichen Li(8), Kohei Sato(3,4), Tristan D. Clemons(3,4,9), Elizabeth L. Daley(1), Thao P. Nguyen(1,10), Eric N. Anderson(7), Justin K. Ichida(8), Udai B. Pandey(7), Sandra Wolin(6), Samuel I. Stupp(3,4,10,11,12) and Evangelos Kiskinis(1,4,13)

31. Combined treatment with nicotinamide riboside, pterostilbene and ibudilast delays motor neuron death in murine models of ALS

Elena Obrador*(1,2), Rosario Salvador-Palmer (1), Paz Moreno-Murciano (2), María Oriol-Caballo (1), Ryan W. Dellinger (3), José M. Estrela (1,2), Rafael López-Blanch (1,2)

32. Contribution of inhibitory neurons to ALS and FTD-like phenotypes linked to FUS

Félicie LORENC (*) (1), Vanessa KAN (2, 3), Luc DUPUIS (1), Sabine LIEBSCHER (2), Raphaelle CASSEL (1)

33. Contribution of one-carbon metabolism to neurodevelopment and neurodegeneration in mouse models of ALS

Marina Hernán Godoy* (1), Geoffrey Stuart-Lopez (1), Caroline Rouaux (1)

34. C-terminal frameshift variant of TDP-43 with pronounced aggregation-propensity causes rimmed vacuole myopathy but not ALS/FTD

Pedro Ervilha Pereira (1, 2)*, Nika Schuermans (1, 2), Antoon Meylemans (3, 4), Pontus LeBlanc (1, 2), Lauren Versluys (1, 2), Katie E. Copley (5, 6), Jack D. Rubien (5), Christopher Altheimer (7), Myra Peetermans (1, 2), Elke Debackere (1, 2), Olivier Vanakker (1, 2), Sandra Janssens (1, 2), Jonathan Baets (8, 9, 10), Kristof Verhoeven (3, 11), Martin Lammens (12), Sofie Symoens (1, 2), Boel De Paepe (3, 4), Sami J. Barmada (7), James Shorter (5, 6), Jan L. De Bleecker (3, 4), Elke Bogaert (1, 2), Bart Dermaut (1, 2)

35. Cyclodextrins As Therapeutic Agents in a poly-GA Mouse Model

Ali Rezaei(*)(1,2,3), Zeynep Günes(3,5), Nellwyn Hagan(4), Franz Bauernschmitt(5), Virag Kocsis-Jutka(1,) Sabine Liebscher(3,5), Eduardo Beltran(5), Dimitry Ofengeim(4), Dieter Edbauer(1,2,3)

36. CYP46A1 as a relevant target to treat ALS pathology independant from its origin

Guillaume Wurtz1*, Haniyeh Yousef Zadeh1*, Emilie Audouard1, Cedric Hermann1*, Antonin Lamaziere2, Nathalie Cartier1*, Françoise PIGUET1

37. Cytokines dysregulation in spinal cord organoids derived from sALS patients

Matteo Bordoni* (1), Eveljn Scarian (1), Maria Garofalo (1), Jessica Garau (1), Francesca Dragoni (1,2), Rosalinda Di Gerlando (1,2), Emanuela Jacchetti (3), Manuela Teresa Raimondi (3), Luca Diamanti (1) Stella Gagliardi (1), Orietta Pansarasa (1)

38. Deciphering the role of Tbk1 in mouse motor neurons and microglial cells, and its implications for ALS pathogenesis.

Lenoël I (1)*, Ribon M (1), Philibert C (1), Lameth J (2), Berriat F (1), Robaldo D (1), Badsi M (1), Gyorgy B (3), Lejeune F-X (3), Messéant J (4), Mallat M (2), Bohl D (1), Millecamps S (1), Brenner D (5), Weishaupt J (6), Boillée S (1), Lobsiger CS (1)

39. Defects in the DNA damage response are Rescued by Enhancing Chromatin Ubiquitination and DDRNAs Biogenesis in ALS Cellular Model System.

F. Esposito(1,2), F. Pisati (3), N. d'Ambrosi (4), M. Cozzolino (5) , F. D'Adda di Fagagna (1,3) and S. Francia (1,3)

40. Defining the role of histone H1.2 in the pathogenesis of FUS associated Amyotrophic Lateral Sclerosis

Hafiza Alirzayeva(*)(1)(2)(3), Rute Loureiro(1), Franziska Hommen(1)(2), Angela Johns(1)(2), David Vilchez(1)(3)(4)

41. Deletion of endothelial TDP-43 impairs retinal angiogenesis and disrupts the vascular barrier in the central nervous system.

Víctor Arribas(1), Yara Onetti(1), Marina Ramiro-Pareta(2,3), Pilar Villacampa(1), Ofelia M. Martínez-Estrada(2,3), Heike Beck(4), Markus Sperandio(4), Bettina Schmid(5), Eloi Montanez(1),*

42. Development of an AAV gene therapy for C9orf72 ALS by targeting the repeat expansion containing C9orf72 transcripts

Vanessa Zancanella, Anna Aster de Ruiter, Romy M van Vliet, Kristel S van Rooijen, Madhurima Chatterjee, Ines Lopes Perreira, Vlad Shtefaniuk, Sebastian N Kieper, Irena Bockaj, Morgane Wartel, Elena de Miguel, Greg Dobrynin, Mercedes Valls Seron, Jovana J Kovačević, *Ying Poi Liu

43. Differential neuronal vulnerability to TDP-43 pathology in a mouse model of amyotrophic lateral sclerosis.
Abedallah Hamida*(1), Rebecca Sneddon(1), Julia Hakkarainen(1), Simon H. Parson(1)

44. Differential pathways of astrocyte toxicity converge in common neurotoxicity markers
Katie Bowden, Chloe Allen, Mark Dunning, Guillaume Hautbergue, John Cooper Knock, Pamela J Shaw and Laura Ferraiuolo.

45. DNA Damage Response Defects are Rescued by Enhancing Chromatin Ubiquitination and DDRNAs Biogenesis in TDP-43 and FUS P-525L ALS Cellular Model System
Francesca Esposito1,2, Stefania Modafferi1,3, Ubaldo Gioia1, Stefania Farina4, Federica Pisati, Nadia d'Ambrosio7, Mauro Cozzolino7, Gianluca Cestra6, Fabrizio D'Adda Di Fagagna5 and Sofia Francia1*

46. Dysregulation of the endocannabinoid system in human post-mortem samples from patients with frontotemporal dementia
Claudia Gonzalo-Consuegra*1-3, Carmen Rodríguez-Cueto1-3, Alberto Rábano4, Javier Fernández-Ruiz1-3 and Eva de Lago1-3

47. Early mechanisms of neurotoxicity associated with intercellular spreading of TDP-43 pathology
Laura Rodríguez-Gómez* (1), Oihane Pikatza-Menoio (1, 2), Maddi Garciandia-Arcelus (1), Andrés Jiménez-Zúñiga (1), Jon Ondaro-Ezkurra (1, 2), Gorka Guereñu-Lopetegi (1, 2), Adolfo López de Munain (1, 2, 3, 4), Sonia Alonso-Martín (1,2), Francisco Javier Gil-Bea (1, 2, 5)

48. Effect of high-fat diet on hippocampal glutamatergic transmission and neuroinflammation in a murine model of amyotrophic lateral sclerosis
Laura Romero-Muñoz*(1), Ana Belén Sanz-Martos (2), Beatriz Merino Palacios (1), Nuria del Olmo (2), Victoria Cano (1), Carmen Fernández-Martos (1).

49. Effect of mutations in the C-terminal region of Tardbp in knock-in mice at the phenotypic and molecular level
José Miguel Brito Armas* (1, 2), Erick Castillo Vargas (2), Judith Noda Mayor (1, 2), Ramón Alejo Muñoz de Bustillo Alfaro (2), Lucas Taoro González (2), Felipe Fumagallo Reading (2), , Elizabeth MC Fisher (3), Pietro Fratta (3), Thomas Cunningham (4) y Abraham Acevedo Arozena (1, 2).

50. Effects of VIP and the ADNP-derived peptide NAP in the SOD1(G93A) mouse model of ALS: physiological and therapeutic significance
Diana Caballero-Hernandez (1), Marta Cejudo-Guillen (1), Juan Polo-Padillo (2), Rafael Fernandez-Montesinos (1), Marien Cobo (3), Francisco Martín (3), Jaime M. Franco (1), Cintia Roodveldt (1,4) David Pozo *(1,4)

51. Efficacy of Sodium Phenylbutyrate and Ursodoxicoltaurine Combination in Transgenic Mice Displaying Progressive Motor Neuron Degeneration Phenotype
*Jamie Timmons (1), Joshua Cohen (1), Justin Klee (1)

52. Elucidating mechanisms underlying FUS aggregation and spreading in neurodegeneration
Britt Tilkin*(1) and Sandrine Da Cruz (1)

53. Elucidating the contribution of the C-terminal sequence of ALS mutated KIF5A in protein aggregation
Pietro Zanella*1, Shreya Agarwal2, Sala Carlo1, Verpelli Chiara1, Tobias Böeckers1, Alberto Catanese2

54. EST79232 and EST79376, two novel Sigma-1 Receptor ligands, exert neuroprotection on models of motoneuron degeneration

Núria Gaja-Capdevila (1)(2)*, Neus Hernández (1)(2), Carla Badia Puiggalí (1)(2), Sandra Yeste (3), Raquel F. Reinoso (3), Javier Burgueño (3), Ana Montero (3), Manuel Merlos (3), José M. Vela (3), Mireia Herrando-Grabulosa (1)(2), Xavier Navarro (1)(2)

55. Excitation/ Inhibition Balance in Amyotrophic Lateral Sclerosis (ALS)

Kareen Halablab (1,2), Natalie Dikwella (1), Oumayma Aousji (1), Stefano Antonucci (1), Diana Wiesner (2), Francesco Roselli (1,2)

56. Gene editing reveals ADNP protein (Activity-Dependent Neuroprotective Protein) physiological roles in microglial and its potential implications in neu

Lucía Silvera-Carrasco *(1,2), Raquel García-García (1,2), Enrico Tebaldi (1), Jesus A. Perez-Cabello, Araceli Rodríguez-Martín, Daniel Tejada-Moreno (1), Alejandro Sola-García (1), Alejandro Martín-Montalvo (1), Xiaoyun Sun (3), Yuhua Sun (3), Cintia Roodveldt (1,2), David Pozo (1,2)

57. GEOexplorer: a webserver for gene expression analysis and visualization

Guy P Hunt*, Luigi Grassi, Rafael Henkin, Fabrizio Smeraldi, Thomas P Spargo, Renata Kabiljo, Sulev Koks, Zina Ibrahim, Richard J B Dobson, Ammar Al-Chalabi, Michael R Barnes, Alfredo Iacoangeli

58. Harnessing Transcriptomic Signals for Amyotrophic Lateral Sclerosis to Identify Novel Drugs and Enhance Risk Prediction

Oliver Pain (*,1), Ashley Jones (1), Ahmad Al Khleifat (1), Devika Agarwal (2), Dzmitry Hramyka (3,4), Hajer Karoui (5), Jędrzej Kubica (6,7), David J. Llewellyn (8,9), Janice M. Ranson (8), Zhi Yao (10), Alfredo Iacoangeli (1,11,12), Ammar Al-Chalabi (1)

59. hnRNPH in nuclear G4C2 foci and cytoplasmic stress granules of C9ORF72 ALS

Nives Škorja Milić(1*), Sonja Prpar Mihevc(1,) Valter Bergant(1,3), Julija Mazej(1), Urša Čerček(1), Alfredo Castello(4), Gregor Gunčar(2), Vera Župunski(2), Boris Rogelj(1,2)

60. Identifying compounds for drug repurposing in ALS using a TDP-43 mutant mESC-derived motor neuron model

Emily Carroll*(1,2), David Gordon (1,2), Stephanie Hatch (3), Helena Chaytow (4), Thomas Gillingwater (4), Daniel Ebner (3), Kevin Talbot (1,2)

61. Identifying Forkhead Box Q1, as a novel regulator of ferroptosis

M. Ismail a,b S. Doll a, B. Proneth a, A. Storch b, U. Walter b, M. Conrad a

62. IFB-088, a multifunctional drug candidate targeting the major ALS cellular pathophysiological mechanisms

Emmanuelle Abgueguen*(1), Massimo Tortarolo(3), Laura Camporeale(3), Caterina Bendotti(3), Alexandre Henriques(2), Noëlle Calizot(2), Philippe Guédat(1), Aurélie Martin(1), Caroline Treins(1), Rozenn Le Bloas(1), Pierre Miniou(1)

63. Imaging the spinal cord neurodegeneration of the TDP-43-A315T ALS mouse model: relationship between MRI and TDP-43 aggregates

Débora Lanznaster* (1), Yara Al Ojaimi (1), Sandra Meme (2), Rudy Clemençon (2), Patrick Vourc'h (1), Hélène Blasco (1).

64. Immune-mediated myogenesis and acetylcholine receptor clustering promote a slow disease progression in ALS mouse models

Cassandra Margotta* (1), Paola Fabbrizio (1), Marco Ceccanti (2), Chiara Cambieri (2), Gabriele Ruffolo (3,4), Jessica

D'Agostino (1), Maria Chiara Trolese (1), Pierangelo Cifelli (5), Veronica Alfano (4), Christian Laurini (2), Silvia Scaricamazza (6), Alberto Ferri (6,7), Gianni Sorarù (8), Eleonora Palma (3,4), Maurizio Inghilleri (2), Caterina Bendotti (1) and Giovanni Nardo (1)

65. Impact of parthenolide on primary microglial cells and motor neurons derived from mutant SOD1-G93A mice

Thomas Gschwendtberger(1)(*), Nadine Thau-Habermann(1), Susanne Petri(1)

66. Impaired cytoplasmic dynein mediated transport of signalling endosomes in motor neurons dysregulates ERK1/2 signalling pathway

Conor McKiernan* (1), Greig Joilin (1), Libby Moody (1), Majid Hafezparast (1)

67. Impaired response to hypoxia in a physiologically relevant Drosophila model of C9orf72 FTD/ALS.

Duncan Garner* (1,2), Ryan J.H. West (1,2)

68. IMPROVING PATHOLOGICAL PROTEIN ANALYSIS BY FLOW CYTOMETRY AND IMMUNOFLUORESCENCE IN ALS.

Paula Fernández-Gómeza, Carlota Tosat-Bitriánb, Ángeles Martín Requerob,c, Valle Palomoa,c.

69. In vivo functional validation of the ALS causative p.E696K missense mutation in TBK1.

Ibrahim Raya (1,2)#, Yasmin Douahem (1)#, David Brenner (1,3), Rüstem Yilmaz (1), Volker Ast (4,5), Angelika Duda (4,5), Qi Gao (6), Riccardo Cristofani (2), Angelo Poletti (2), Vittorio Maglione (7), Jochen Weishaupt (1)§, Rosanna Parlato (1)§*

#, § equal contributions

70. "Increased ADAM 10/17 activity in an animal model of ALS: rationale for targeting ADAMs as potential therapeutic target?"

Cabras Paolo(1)*, Spatafora Mauro G.(1), Dimartino Agnese(1), Teodoro Manilla(1), Gazzano Andrea(1) , Peviani Marco(1)#.

71. Insights into KIF5A-related pathways to neurodegeneration

Marta Cozzi (1)(*), Barbara Tedesco (1), Veronica Ferrari (1), Elena Casarotto (1), Marta Chierichetti (1), Paola Pramaggiore (1), Guglielmo Patelli (1), Margherita Piccolella (1), Mariarita Galbiati (1), Paola Rusmini (1), Valeria Crippa (1), Cinzia Gellera (2), Daniela Di Bella (2), Stefania Magri (2), Riccardo Cristofani (1), Franco Taroni (2), Angelo Poletti (1)

72. Integrative proteomics highlight presynaptic alterations and c-Jun misactivation as convergent pathomechanisms in ALS

Amr Aly* (1), Tobias M. Böckers (1)(2), Alberto Catanese (1)(2)

73. Interactomes of wt and C-terminally truncated FUS

Helena Motaln (1), Miha Milek (2), Katarina van Midden (1), Boris Rogelj (1,3*)

74. Intracerebroventricular delivery of AAV9-based gene therapy selectively transduces motor neurons in the spinal cord when delivered to neonatal mice

Alannah J Mole (1)*, Amisha R Parmar (2), Amy FA Keerie (1), Richard J Mead (1).

75. Investigating Purine Metabolism in C9orf72 ALS

Benjamin P. C. Hall*, Jonathan G. George, Keaton Hamer, Guillaume M. Hautbergue, Adrian Higginbottom, Kurt J. De Vos and Scott P. Allen.

76. Investigating the Role of NEK1 in ALS.
Natalie Pye* (1), Andrew Grierson (1), Kurt De Vos (1).

77. INVESTIGATING VARIANTS IN NUP50 AS RISK FACTORS FOR AMYOTROPHIC LATERAL SCLEROSIS
Olga Roman* (1), Salim Megat (1), Chantal Sellier (1), Luc Dupuis (1)

78. LncRNAs in muscle of SOD1G93A ALS model mice: potential as biomarkers and intervention targets.
Tresa López-Royo (*) (1), Laura Moreno-Martínez (1), Leticia Moreno-García, Ana Cristina Calvo, Alberto García-Redondo (2), Raquel Manzano (1), Rosario Osta (1).

79. Loss of CHK1 contributes to DNA damage accumulation in cells bearing WT TDP-43 and FUS P525L cytoplasmic inclusions
Stefania Modafferi(1,2*), Francesca Esposito(1,3), Stefania Farina(1), Ubaldo Gioia(1,4), Fabrizio D'Adda Di Fagagna(1,4), Sofia Francia(1,4)

80. Loss of HSF1 and HSC70 protection in spinal motor neurons contributes to the onset of ALS pathogenesis
Vadreenath Tripathy (1), Priyanka Tripathi (2), Ramakrishnan Sivasubramanian (1), Tyler R. Fortuna (3), Amay A. Agrawal (1), Jessica Bellmann (1), Lara Marrone (1), Dimitra Alexopoulou (4), Marco Cimino (5), Andreas Dahl (4), Dirk Troost (6), Eleonora Aronica (6), Serena Carra (5), Joachim Weis (2), Udai Bhan Pandey (3,7,8), Anand Goswami (2), Jared Sterneckert* (1)

81. MAPK/MAK/MRK overlapping kinase (MOK) regulates microglial inflammatory/type-I IFN responses via Brd4 and is involved in ALS
Jesús A. Pérez-Cabello (1,2), Jaime M. Franco (1), Lucía Silvera-Carrasco (1,2), Vivian Capilla-González (1), Alexandros Armaos (3), María Gómez-Lima (1), Raquel García-García (1,2), Xin Wen Yap (4), M. Magdalena Leal-Lasarte (1), Deepti Lall (5), Robert H. Baloh (5), Salvador Martínez (6), Yoshihiko Miyata (7), Gian G. Tartaglia (3,8), Ritwick Sawarkar (4), Mario García-Dominguez (1), David Pozo (1,2), Cintia Roodveldt (1,2)*

82. MCH neurons in ALS: vulnerability and connectivity
Jelena Scekic-Zahirovic (1*), Diana Wiesner (1), Stefano Antonucci (2), Chiara Ebner (4), Hussein El Hajj (2), Rosa Depau (4), Gizem Yartas (1), David Bayer (3) Amela Londo (2), Albert C. Ludolph (1,2), Francesco Roselli (1, 2)

83. Metabolic profiling of fibroblasts derived from healthy donors and Amyotrophic Lateral Sclerosis patients bearing the p.G376D mutation in the TARDBP g
Elisa Perciballi (1), Federica Bovio (1), Ivan Lombardi (1) J. Rosati (2), A. D'Anzi (2), Serena Lattante (3), Mario Sabatelli (3), Vincenzo La Bella (4), Viviana Mosca (4), Tarja Malm (5), Angelo Vescovi (1), Paola Fusi (1), Daniela Ferrari (1)

84. Mitochondrial genome study in blood of maternally-inherited ALS cases
Sarah J. Brockmann (1)*#, Eva Buck (2)#, Tiziana Casoli (3), João L. Meirelles (2), Wolfgang P. Ruf (1), Paolo Fabbietti (4), Karlheinz Holzmann (5), Jochen H. Weishaupt (1,6), Albert C. Ludolph (1,2), Fiorenzo Conti (3, 7) and Karin M. Danzer (1, 2) # These authors contributed equally

85. Mitochondrial TSPO overactivation via ERK1/2 cascade correlates with impairment of respiration and mitophagy in a mouse model of ALS
Andrea Magrì (1)*, Cristiana Lucia Rita Lipari (2), Giuseppe Battiato (2)
Francesca Guarino (2), Antonella Caccamo (3), Angela Messina (1)

86. Modeling C9ORF72 DPR pathology using optogenetics.

Jessica Rayment (1), Rachel Hodgson (1), Tatyana Shelkovnikova (1)

87. Modulating the gut dysbiosis by dietary intervention in a murine model of ALS.

Leticia Moreno-García(1), Inés Valledor Martín(1), Laura Moreno Martínez(1), Janne Markus Toivonen(1), Ana Cristina Calvo(1) (*), Rosario Osta(1)

88. Mono-methylation of arginine 293 of TDP-43 and its function in translational control

Sarah Müller 1, Katharina Limm 2, Sarah J. Brockmann 1, Lea-Marie Mauder 1, Lorena Decker 1, Sonja Menge 1, Marco Zimmel 1, Peter J. Oefner 2, Albert C. Ludolph 1, 3, Jochen H. Weishaupt 4, Axel Freischmidt 1

89. Neurofilament accumulations in Amyotrophic Lateral Sclerosis patients' motor neurons impair axonal initial segment integrity.

Cynthia Lefebvre-Omar (1), Elise Liu (1), Carine Dalle (1), Boris Lamotte d'Incamps (2), Stéphanie Bigou (1), Clément Daube (1), Marc Davenne (1), Noémie Robil (3), Léa Karpf (1), Coline Jost Mousseau (1), Stéphane Blanchard (4), Guillaume Tournaire (4), Charles Nicaise (5), François Salachas (1, 6), Lucette Lacomblez (6), Danielle Seilhean (1, 7), Christian S. Lobsiger (1), Stéphanie Millecamps (1), Séverine Boillée (1), Delphine Bohl* (1).

90. Neuroimmune characterization of aged mice with optineurin insufficiency

Josip Peradinovic* (1), Nikolina Mohovic (1), Andrea Markovinovic (2), and Ivana Munitic(1)

91. Neuromuscular junction denervation and terminal Schwann cell loss in the hTDP-43 overexpression mouse model of amyotrophic lateral sclerosis (ALS)

Hannah L. Smith* (1, 2), Abrar Alhindi (1, 2, 3), Megan Shand (1, 2), Ana S. Leite (1, 2, 4), Yu-Ting Huang (1, 2), Dinja van der Hoorn (1, 2), Zara Ridgway (1, 2), Kiterie M.E. Faller (5), Ross A. Jones (1, 2), Thomas H. Gillingwater (1, 2), Helena Chaytow (1, 2)

92. Neuromuscular ultrasound as preclinical diagnostic biomarker in the SOD1(G93A) mouse model of amyotrophic lateral sclerosis

Camilla Wohnrade(1)*, Nadine Thau-Habermann(1), Thomas Gschwendtberger(1), Julia Rückold(1), Zhong Huang(2), Kirsten Haastert-Talini(2), Susanne Petri(1)

93. No title

Sonja Menge* (1), Lorena Decker (1), Sarah Müller (1), Albert C. Ludolph (1,2), Patrick Oeckl (1,2), Axel Freischmidt (1)

94. Nonclinical evaluation of the efficacy of Neural Stem Cells intracerebroventricular transplant as a possible treatment for ALS

Ivan Lombardi (1, 2)*, Edvige Vulcano (2), Silvia de la Morena (3, 4), Daniela M Rasà (3, 4), Clelia Ferrero(3, 4), Diego Pastor Campos (5), Rose Mary Carletti (6), Maurizio Gelati (7), Daniela Celeste Profico (7), Valentina Grespi (8), Gianmarco Muzi (8), Jessica Rosati (9), Elisa Perciballi (2), Letizia Mazzini (10), Fabiola De Marchi (10), Alessandro Vercelli (3, 4), Salvador Martinez Perez (11), Angelo L Vescovi (2, 6), Marina Boido (3, 4) and Daniela Ferrari (2)

95. Noradrenaline deficiency as a driver of cortical hyperexcitability in amyotrophic lateral sclerosis

Jelena Scekic-Zahirovic (1,2,12*), Aurore Brunet (1,12), XiaoQian Ye (3,4) , Evgeny Logunov (3,4) , Vincent Douchamp (5), Salim Megat (1), Virginie Andry (6), Vanessa Wing Yin Kan (3,4,7), Geoffrey Stuart-Lopez (1), Johan Gilet (1), Margaux Trombini (1), Stéphane Dieterle (1), Jérôme Sinniger (1), Mathieu Fischer (1), Frédérique René (1), Zeynep Gunes (3,4,7), Pascal Kessler (8), Luc Dupuis (1), The Neuro-CEB Neuropathology Network (9), Yannick Goumon (6), Romain Goutagny (5), , Sabine Liebscher (4,7,10,,11) & Caroline Rouaux (1,11)

96. Novel functionalized nanoparticles targeted to 18KDa translocator protein (TSPO) to track and modulate neuroinflammation in animal models of fALS

Gazzano A.1*, Davide C.1, Spatafora M.G.1, Lamacchia A.1, Marsala A. 1,Stanchina A. 1, Doria E.2, Sponchioni M.3, Auriemma R.3, Lasciafari A.4, Filibian M.4, Moscatelli D.3, Peviani M.1#

97. Novel humanised conditional mouse models of SOD1-ALS

David Thompson (1)*, Georgia Price (1), Jonathan D Gilthorpe (3), Chloe Williams (3), Charlotte Tibbit (1), Rosie Bunton-Stasyshyn (1), Anny Devoy (2), Alasdair J Allan (1), Gemma F Codner (1), Lydia Teboul (1), Jackie Harrison (1), Gemma Atkins (1), Ben Davies (5), Elizabeth M C Fisher (1,2), Thomas J Cunningham (1,4)

98. Novel insights on the role and therapeutic potential of Glycoprotein nonmetastatic melanoma protein B (Gpnmb) in Amyotrophic Lateral Sclerosis.

Mauro G. Spatafora(1)*, Paolo Cabras(1), Gianluca Di Nolfi(1), Andrea Gazzano(1), Teodoro Manilla(1), Loris Bandirali(1), Bruno M Custode(1), Agnese Dimartino(1), Daniela Curti(1), Alessandra Biffi(2), Teuta Domi(3), Nilo Riva(4), Marco Peviani(1)#

99. Novel nuclear bodies bridge FUS autoregulation and phase separation and are disrupted by ALS mutations

Vedanth Kumar(*)(1), Wang-Ping Huang (1), Haiyan An (2), Tatyana Shelkovnikova(1)

100. Novel transcriptional and mutational signatures in amyotrophic lateral sclerosis revealed by multiomics and machine-learning

Dr. Alberto Catanese(1;2)*, Prof. Medhanie Mulaw(3)

101. Nucleocytoplasmic transport factors as modifiers of TDP-43 proteinopathy

Bilal Khalil (1), Deepak Chhangani (2), Melissa C Wren (1), Courtney L Smith (1,3), Jannifer H Lee (1 3,) Xingli Li (4), Christian Puttinger (1), Chih-Wei Tsai (1), Gael Fortin (1), Dmytro Morderer (1), Junli Gao (1), Feilin Liu (1), Ankur Gadgil (1), Chun Kim Lim (5), Jingjiao Chen (1,6), Ching-Chieh Chou (7), Cara L Croft (8,9), Amanda M Gleixner (10,11), Christopher J Donnelly (10,11), Todd E Golde (2,8), Leonard Petrucelli (1), Björn Oskarsson (12), Dennis W Dickson (1), Ke Zhang (1), James Shorter (13), Shige H Yoshimura (5), Sami J Barmada (4), Diego E Rincon-Limas (2,8,14), Wilfried Rossoll* (1)

102. OMIC characterization of patient-derived spinal cord organoids to assess novel genes associated with C9orf72-Amyotrophic Lateral Sclerosis

Noemi Galli* (1), Mafalda Rizzuti (2), Jessica Ongaro (2), Linda Ottoboni (2), Valentina Melzi (2), Monica Nizzardo (2), Giacomo Pietro Comi (1, 2), Stefania Corti (1, 2)

103. Phenotype changes of microglia in SOD1G93A mice after mGluR5 genetic down regulation.

Tiziana Bonifacino (1*), Matilde Balbi (1), Silvia Ravera (2), Daniela Fenoglio (3,4), Tiziana Altosole (3), Riccardo Leardi (5), Giambattista Bonanno (1,6) Marco Milanese (1,4)

104. Phosphorylated Tyr526 FUS in FTD

Helena Motaln (1*), Urša Čerček (1), Anand Goswami (2), Boris Rogelj (1,3)

105. Presence of stathmin-2 cryptic exon alone is not sufficient to determine clinical phenotype in amyotrophic lateral sclerosis/frontotemporal dementia

Fergal M. Waldron (1), Anna-Leigh Brown (2), Olivia M. Rifai (3), Judi O'Shaughnessy (4), Mathew H. Horrocks (4), Pietro Fratta (2), Jenna M. Gregory (1*).

106. Pridopidine exerts neuroprotective effects via activation of the Sigma-1 receptor (S1R)

Randal Hand(1)(*), Lior Ankol(2), Ariel Ionescu(2), Tal Gradus-Pery(2), Shao-Ming Wang(3,4), Noga Gershoni-Emek(1), Tsung-Ping Su(5), Eran Perlson(2), Michal Geva(1), and Michael R. Hayden(1,6)

107. Propagation of SOD1 pathologic determinants in human iPSC-derived motor neurons.

Coline Jost Mousseau*(1), Karpf Léa (1), Gaïzka Le Goff (1), Alexandre Mezghrani (2), Flavien Picard (3), Pascal Leblanc (3), Raoul Cédric (2), Christian S Lobsiger (1), Stéphanie Millecamps (1), Séverine Boillée (1), Delphine Bohl (1)

108. Propranolol reduces the accumulation of cytotoxic aggregates in C9orf72-ALS/FTD in vitro models

Mira Seidel (1)*, Sandeep Rajkumar (1), Christina Steffke (1), Shreya Agarwal (1,2), Albert Ludolph (3,4), Tobias Boeckers (1,4), Alberto Catanese (1,4)

109. Proteomic Changes following Terazosin Treatment in a TDP-43 Mouse Model

Helena Chaytow* (1,2), Rachel Kline (1,3), Dinja van der Hoorn (1,2), Yu-Ting Huang (1,2), Thomas Wishart (1,3), Douglas Lamont (4), Thomas H. Gillingwater (1,2)

110. Regulation of stress granules' formation with plant extracts containing polyphenols

Vera Župunski (1)(*), Jerneja Nimac (2), Lea Pogačnik da Silva (3), Boris Rogelj (1,2)

111. Reprogrammed astrocytes from a C9-ALS family with variable penetrance display differential C9orf72 pathology and motor neuron toxicity in co-culture

Allan C. Shaw* (1), Iris S. Pasniceanu (1), Matthew Wyles (2), Dr Matthew Parker (3,4), Dr Adrian Higginbottom (1,5), Dr Matthew R. Livesey (1,5), Professor Guillaume M. Hautbergue (1,5), Professor Laura Ferraiuolo^ (1,5), Professor Dame Pamela J. Shaw^ (1,5). ^These authors contributed equally to this work.

112. Resistant and vulnerable motor neurons show unique temporal gene regulation in SOD1G93A ALS

Irene Mei (1*), Susanne Nichterwitz (1), Melanie Leboeuf (1), Jik Nijssen (2), Christoph Schweingruber (1,2), Eva Hedlund (1,2)

113. RIPK1 activation contributes to ALS pathophysiology and drives a pro-inflammatory gene signature in a human iPSC-derived tri-culture system

Matija Zelic (1), Anna Blazier (1), Fabrizio Pontarelli (1), Michael LaMorte (1), Ozge E. Tasdemir-Yilmaz (1), Tracie Treleaven (1), Emma McGuirk (2), Yi Ren (1), Jim Dodge (1), Nellwyn Hagan (1), Nazem Atassi* (1), Joseph Gans (1), Giorgio Gaglia (1), Timothy R. Hammond (1), Dimitry Ofengeim (1)

114. Role of MOK kinase in microglial TBK1/type-1 IFN pathway and mitochondrial function

Raquel García-García (1,2), Lucía Silvera-Carrasco (1,2), Enrico Tebaldi (1), Araceli Rodríguez-Martín (1), Daniel Tejada-Moreno (1), Alejandro Sola-García (1), Alejandro Martín-Montalvo (1), David Pozo (1,2), Cintia Roodveldt (1,2)*

115. Simultaneous activation of two complementary targets, Kv7.2/3 and TSPO: a promising and novel treatment for Amyotrophic Lateral Sclerosis

Vera M.Masegosa*(1), Elsa Fritz(2), Daniela Corvalan(2), Fabiola Rojas(2), Xavier Navarro(1), Brigitte van Zundert(2), Mireia Herrando-Grabulosa(1)

116. Single cell RNA sequencing in isogenic FUS and TARDBP mutant ALS lines reveals early mitochondrial dysfunction as a common pathway in motor neurons

Christoph Schweingruber (*,1,2), Jik Nijssen (2), Jonas Mechtersheimer (3), Stefan Reber (3), Michaela Keuper (4), Irene Mei (1), Julio Aguila Benitez (2), Niamh O'Brien (3), Martin Jastroch (4), Marc-David Ruepp (3,5), Eva Hedlund (1,2,5)

117. Single-cell RNAseq in Drosophila models of ALS to decipher non-cell autonomous mechanisms of neurodegeneration

Andrés Jiménez (1,2,*), Gorka Gereñu (1,2,3), Francisco Javier Gil (1,2,4,5), Adolfo López de Munain (1,2,6,7,8), José Luis Zúñiga (1), Jon Ondaro (1,2), Laura Rodríguez (1), Maddi Garciandía (1), Haizea Hernández (1), Saioa Moragón (1,2), Ángela Sánchez (1,9)

118. SOD1-G93A mutation modifies cellular properties of mesenchymal stem cells in a mouse model of Amyotrophic Lateral Sclerosis

Amaya Rando*(1, 2, 3), Paula Aparicio**(1, 2, 3), Laura Barrachina (1, 3), Alina Cequier (1, 3), Ana Rosa Remacha (4), Raquel Manzano (1, 2, 3), Clementina Rodellar (1, 3), Rosario Osta (1, 2, 3) .

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119. Strain-specific propagation of templated SOD1 aggregation along neuroanatomical tracts in ALS mice

Isil Keskin* (1), Elaheh Ekhtiari Bidhendi (1), Matthew Marklund (1), Peter M. Andersen (2), Stefan L. Marklund (1), Thomas Brännström (1), Ulrika Nordström (2)

120. Structure and function of the C9ORF72-SMCR8-WDR41 complex and its implication in Amyotrophic lateral Sclerosis (

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121. TDP-43 dependent splicing missregulation in ALS

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Brittany Ellis* (1), Tatyana Shelkovnikova (1)

123. TDP-43 mutation is associated to intestinal inflammatory phenotypes

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Ruxandra Dafinca1,2*, Carlota Tosat-Bitrian3, Jakub Scaber1,2, Kevin Talbot1,2

125. TDP-43 splicing function and disease evolution in the FTD-ALS spectrum: Insights from a transgenic mouse model

Pascual Torres (1)*, Victoria Ayala(1), Reinald Pamplona(1), and Manuel Portero-Otín (1)

126. The cell autonomous effects on sporadic ALS onset and survival

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Nikki S. Harper *(1), Andreas Prokop (2), Ryan J.H. West (3,4)

128. The Role of Cystatin C in Amyotrophic Lateral Sclerosis

Sarah Granger (*)(1), Prof Kurt De Vos (1), Dr J Robin Highley (1)

129. The Role of Microglia-released microRNAs in ALS Pathology

Libby Moody* (1), Eleni Christoforidou (1), Greig Joilin (1), Sarah Newbury (2), Nigel Leigh (2), Lisa Mullen (2), Majid Hafezparast (1).

130. The role of Serum Response Factor (SRF) in the regulation of autophagy

Natalie Dikwella* (1), (2), #, Jialei Song (1), (2), #, Daniela Sinske (1), Francesco Roselli (2) (3) , Bernd Knöll (1)

 # both authors equally contributed

131. Therapeutic potential of parthenolide in the SOD1-G93A mouse model of Amyotrophic Lateral Sclerosis?

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132. THERAPEUTIC POTENTIAL OF THE NRF2 ACTIVATOR DIMETHYL FUMARATE IN TDP-43-RELATED FRONTOTEMPORAL DEMENTIA MOUSE MODEL

Raquel Martín Baquero (1-3*), Ignacio Silva-Llanes (1,4*), Alicia Berrojo-Armisen (4), Carmen Rodríguez-Cueto (1-3), Javier Fernández-Ruiz (1-3), Eva De Lago (1-3), Isabel Lastres-Becker (3,4)

133. Transcriptomics Analyses of ALS Postmortem Motor Cortex highlight alteration and potential biomarkers in the Neuropeptide Signalling pathway

Renata Kabiljo(1), Heather Marriott(1,2), Guy P Hunt(1,2,3,4), Abigail L Pfaff(3,4), Ahmad Al Khleifat(2), Brett Adey(1,9), Ashley Jones(2), Claire Troakes(10), John P Quinn(5), Richard J B Dobson(1,7,8), Sulev Koks(3,4), Ammar Al-Chalabi(2,6), Alfredo Iacoangeli(1,2,8)

134. U1 snRNA as a novel RNA-based therapeutic approach to modulate C9ORF72 pathology in patient-derived iPSC-motoneurons

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135. Ultra-high spatial resolution ex-vivo structural brain MRI reveals early caudoputamen atrophy in FusΔNLS/+ mice

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136. Ultrastructural Imaging of Distinct SOD1 Strains in ALS Transgenic Mouse Models

Isabelle Sigfridsson*(1), Thomas Brännström (1)

137. Unconventional secretion of misfolded SOD1 and toxicity spreading: a novel therapeutic strategy for ALS

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138. UNCONVENTIONAL SECRETION OF TDP-43 FREE AGGREGATES BY THE UBIQUITIN-SPECIFIC PROTEASE 19 (USP19)

Flavien PICARD* (1), Elisabeth ERRAZURIZ-CERDA (2), Edwige BELOTTI (1), Takashi NONAKA (3), Coline JOST-MOUSSEAU (4), Valérie RISSON (1), Alexis OSSENI (1), Emilien BERNARD (1, 5), Delphine BOHL (4), Thierry GALLI (6), Laurent SCHAEFFER (1), Cédric RAOUL (7) and Pascal LEBLANC (1)

139. Uncovering the protein interactions of MATR3 and its ALS-associated mutant MATR3 S85C

Jerneja Nimac (1,2*), Anand Goswami (3), Vera Župunski (4), Boris Rogelj (1,4)

140. Uncovering the proteome of TDP-43 condensates in human neurons

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141. Understanding muscle impairment in Amyotrophic Lateral Sclerosis in the absence of motoneuron input

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142. Understanding the neuropathological consequences of NEK1 mutations in ALS

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143. Unrevealling the molecular mechanisms involved in the protection exerted by genetic leptin haploinsufficiency in the mouse model of ALS SOD1G93A

Luis Fernandez-Beltran (1,2), Zeinab Ali (2), Jorge Matias-Guiu Guia (2), Silvia Corrochano * (2).

144. Using optogenetics to model activity-dependent neurodegeneration in amyotrophic lateral sclerosis

Lucy Farrimond* (1), Ruxandra Dafinca (1), Colin Akerman (2), Kevin Talbot (1).

145. Utreloxastat, a Novel 15-Lipoxygenase Inhibitor, Reduces Ferroptosis-Induced Injury for ALS and Related Neurodegenerative Diseases

Angela Minnella, Beatrice Trias, Monal Dieterich, Kelli DesRochers, Kevin McCusker, Lovena David, Yuting Mao, Grace Liu, Meenu Pillai, Xin Zhao, Akiko Amagata, Joey Latham, Marla Weetall, Jeff Trimmer

146. Validation and characterisation of the therapeutic effect of HDAC6 inhibition using a zebrafish model for ALS

Dufie Strubbe (1)(2)(*), Valérie Bercier (1)(2), Julie De Schutter (1)(2), Kara Heeren (1)(2), Philip Van Damme (1)(2)(3) and Ludo Van Den Bosch (1)(2)

147. Validation of Exportin-1 (XPO-1) and Mitogen-Activated Protein Kinase Kinase 2 (MAP2K2) as Molecular Drug Targets in Amyotrophic Lateral Sclerosis

Mojan Parvaz*(1), Lucas Caldi Gomes(1), Sonja Hänzelmann, Sergio Oller(2), Fabian Hausmann(2), Robin Khatri(2), Melanie Ebbing(2), Constantin Holzapfel(2), Johanna Knöferle(1), Isabell Cordts(1), Antonia F. Demleitner(1), Laura Tzeplaeff(1), Marcus Deschauer(1), Marc Sturm(3), Tobias Haack(3), Sebastian Streb(4), Magdalena Kuzma-Kozakiewicz(5), Marie Gebelin(6), Christine Carapito(6), Pavol Zelina(7), Marta Canizares Luna(7), R. Jeroen Pasterkamp(7), Laura Pasetto(8), Valentina Bonetto(8), Qihui Zhou(9), Dieter Edbauer(9), Endre Laczko(4), Hubert Rehrauer(4), Ralph Schlapbach(4), Stefan Bonn(2), Paul Lingor(1).

148. Whole-genome bisulfite sequencing of motor neurons reveals cell specific enhancers & enables the identification of motor neuron derived cell free DNA

Calum Harvey *(1), Eilis Hannon (2), Annika K Weimer (3), Christa Caggiano (4,5), Minyi Shi (3), Tobias Moll (1), Cleide Dos Santos Souza (1), Sai Zhang (3,6), Ke Ning (1), Aina Barcons (1), Kevin P Kenna (7), Ilia Timpanaro (7), Charlotte van Dijk (7), Noah Zaitlen (4,8), Laura Ferraiuolo (1), John Mills (2), Michael Snyder (3), Pamela Shaw (1), Jonathan Cooper-Knock (1)

149. WWOX contributes to mitochondrial dysfunction and oxidative stress in ALS

Tiziana Petrozziello (1)(*), Spencer E. Kim (1), Alexandra N. Mills (1), Sommer S. Huntress (1), Ayleen L. Castillo Torres (1), Ranee Zara Bunag Monsanto (1), Sali M.K. Farhan (2,3,4), Khashayar Vakili (5), Merit E. Cudkowicz (1), James D. Berry (1), Ghazaleh Sadri-Vakili (1)

150. ZEB1-AS1 IS IMPLICATED IN SPORADIC AMYOTROPHIC LATERAL SCLEROSIS THROUGH BETA-CATENIN MODULATION

Federica Rey (1)(*), Erika Maghraby (1,2), Letizia Messa (3,4), Letizia Esposito(1), Bianca Barzaghini (5), Cecilia Pandini (6), Matteo Bordoni (7), Stella Gagliardi (8), Luca Diamanti (9), Manuela Teresa Raimondi (5), Gianvincenzo Zuccotti (1,10), Stephana Carelli (1,3), Cristina Cereda(3)

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151. 5'Gly-GCC tiRNA levels as a biomarker of disease progression in Amyotrophic Lateral Sclerosis

Koen C Demaegd (1)*, Lindy Kool, (2,3), Hesham A Gibriel (4), Paul Donovan (4), Grainne Geoghegan (4), Elisabeth Jirstrom (4), Luise Halang (4), Philip Van Damme (5), Pegah Masrori (5), Morten Veno (6), Jorgen Kjems (6), Orla Hardiman (7), Leonard H van den Berg (1), Jan H Veldink (1), Kevin Kenna (3), Jochen HM Prehn (4)* & Michael A van Es (1)

152. Biofluid extracellular vesicle extraction and proteomic profiling in ALS

Elizabeth Dellar* (1), Ruxandra Dafinca (1,2), Kevin Talbot (1,2), Martin R Turner (1), Alexander G Thompson (1).

153. Biomarkers of neurodegeneration and muscle wasting in amyotrophic lateral sclerosis – a monocentric cross-sectional study

Maximilian Vidovic (1)*, René Günther (2)

154. Brain 2-[18F]FDG-PET as a tool to discriminate Amyotrophic Lateral Sclerosis (ALS) from ALS-mimics

Sara Cabras* (1,2), Antonio Canosa (1,3,4), Alessio Martino (5), Alessandro Giuliani (6), Cristina Moglia (1,3), Rosario Vasta (1), Maurizio Grassano (1), Francesca Di Pede (1), Paolina Salamone (1), Giulia Marchese (1), Federico Casale (1), Giulia Polverari (7), Marco Pagani (4,8), Umberto Manera (1,3), Andrea Calvo (1,3,9), Adriano Chiò (1,3,4,9)

155. Cell free miRNA biomarkers for ALS prognostication

Iddo Magen* (1), Nancy Yacovzada (1), Yahel Cohen (1), Joanne Wuu (2), Michael Benatar (2), Andrea Malaspina (3), Pietro Fratta (3), Eran Hornstein (1)

156. CHARACTERIZATION OF BLOOD-DERIVED MACROPHAGES OF AMYOTROPHIC LATERAL SCLEROSIS (ALS) FOR THERAPEUTIC AND DIAGNOSTIC APPROACH.

Matthieu Ribon * (1), Adèle Hesters(1)(2), Maria Del Mar Amador(2), Lorraine Bavelier (1), Aude Chiot (1), Gaëlle Bruneteau (2), Timothée Lenglet (2), Philippe Couratier (3) Stéphanie Millecamps (1), Delphine Bohl (1), Christian S Lobsiger (1), François Salachas (1)(2), Séverine Boillée (1).

157. Combined spirometric, arterial blood gas analysis and overnight oximetric parameters for the prognosis of patients affected by motor neuron disease.

Paride Schito,* 1,2 Umberto Manera,3 Tommaso Russo, 1,2 Sara Lavelli,4 George Cremona,4 Andrea Tettamanti,4 Andrea Calvo,3 Federica Agosta,5 Adriano Chiò,3 Nilo Riva,2,6 Massimo Filippi 1,2,5

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158. Cortical and subcortical grey matter atrophy in Amyotrophic Lateral Sclerosis correlates with measures of disease accumulation independent of disease

Nora Dieckmann (1)*, Annekathrin Roediger (1), Tino Prell (2), Simon Schuster (3), Meret Herdick (3), Thomas E. Mayer (4), Otto W. Witte (1), Julian Grosskreutz (3), Robert Steinbach (1)

159. Deciphering the role of non-coding variants in the etiology of neurodegenerative diseases (NDDs) by massively parallel reporter assay (MPRA)

Lucia Corrado1, Beatrice Piola*(1), Fjorilda Caushi(1), Endri Visha(1), Nadia Barizzone(1),Erica Melone(1),Diego Cotella(1), Laura Follia(1),Martina Tosi(1), Fabiola De Marchi(2), Luca Magistrelli(3), Letizia Mazzini(2), Alfredo Brusco(4), Sandra D'alfonso(1).

160. Designing an ALS biomarker panel

Laura Expósito Blázquez (1), Daniel Borrego Hernández (1), Janne Markus Toivonen (2), Ana Cristina Calvo (2), María del Carmen Herrero Manso (3), Pilar Cordero Vázquez (1), Alberto Villarejo Galende (4), Sara Llamas Velasco (4), Marta González Sánchez (4), Miguel Ángel Martín Casanueva (5), Jesús Esteban Pérez (1), Rosario Osta (2) and Alberto García Redondo (1)

161. DISCOVERY OF ACI-19278, A POTENTIAL FIRST IN CLASS TDP-43 PET BIOMARKER FOR FTLD-TDP AND ALS

Elodie Chevalier (*)(1), Tariq Afroz (1), Efthymia Vokali (1), Nicolas Dreyfus (1), Mathieu Clavel (1), Monisha Ratnam (1), Tania Melly (1), Thomas Jaquier (1), Heiko Kroth (1), Andrea Pfeifer (1), Marie Kosco-Vilbois (1), Tamara Seredenina (1)

162. Evaluating the characteristics of High Frequency Evoked Responses as a potential biomarker for sensorimotor dysfunction in ALS: A Preliminary Study

Prabhav Mehra*(1), Saroj Bista(1), Marjorie Metzger(1), Serena Plaitano(1), Rosie Giglia(1), Matthew Mitchell(1), Peter Bede(1), Madeleine Lowery(4), Muthuraman Muthuraman(2), Richard Carson(5), Lara McManus(1), Orla Hardiman(1,3), Bahman Nasseroleslami(1)

163. Evaluation of arterial blood gas parameters as prognostic markers in amyotrophic lateral sclerosis

Hugo Alarcan*# 1 2 , Camille Cotet # 1 , Nadège Lépine 1 , Julie Morel 1 , Patrick Vourc'h 1 2 , Christian Andres 1 2 , Philippe Corcia 2 3 , Hélène Blasco

164. Extracellular Vesicles as Potential Biomarkers in Amyotrophic Lateral Sclerosis

Maruša Barbo*(1), Metka Ravnik Glavač(1)

165. First evidence of altered Gas6-Axl signaling and correlation between sAXL blood levels and clinical decline in ALS

Mauro G Spatafora(1), Paolo Cabras(1), Andrea Gazzano(1), Gianluca Di Nolfi(1), Loris Bandirali(1), Agnese Dimartino(1), Davide Camazzola(1), Danilo Pellin(2), Daniela Curti(1), Alessandra Biffi(2), Teuta Domi(3), Koen Poesen (4), Nilo Riva(5), Marco Peviani(1)(*)

166. Hsa_circ_0060762 and CSE1L are potential peripheral blood biomarkers and therapeutic targets for ALS

Metka Ravnik Glavač (1)*, Massimo Mezzavilla (2) , Ana Dolinar (3) , Blaž Koritnik (4,5), Damjan Glavač (3,6)

167. Hypothalamic correlates of metabolism and cognition in ALS

Annebelle Michielsen* (1), Kevin van Veenhuijzen (1), Mark R Janse van Mantgem (1), Ruben van Eijk (1), Henk-Jan Westeneng (1), Leonard H. van den Berg (1).

168. Identification and validation of a protein biomarker signature from tear fluid in patients with Amyotrophic Lateral Sclerosis (ALS)

Lena-Sophie Scholl* (1), Antonia Franziska Demleitner(1), Jenny Riedel (2,3), Clara Meijs (2,3), Lucas Caldi Gomes(1), Ana Galhoz (2,3), Isabell Cordts (1), Christof Lenz (5,6), Michael Menden (2,3,4),Paul Lingor (1,7,8)

169. Identification of a disease signature for premotor and early ALS

Laura Tzeplaeff (1)(*), Lucas Caldi Gomes (1), Nicholas Ashton (2), Michael Benatar (3), Philippe Corcia (4), Florian Kohlmayer (5), Wojciech Kuban (6), Christof Lenz (7), Yossi Lerner (8), Michael Menden (9), Amrei Menzel (5), Peter Munch Andersen (10), Ana Paulo Galhoz (9), Mary-Louise Rogers (11), Stephanie Shepheard (11), Hilmi Uysal (12), Markus Weber (13), Joanne Wuu (3), Norbert Zilka (14), and Paul Lingor (1).

170. Identification of non-coding RNA that correlate with prognosis and progression in the serum of people with amyotrophic lateral sclerosis.

Greig Joilin* (1), Andre Altmann (2), P Nigel Leigh (3), Janine Kirby (4), Pamela J Shaw (4), Majid Hafezparast (1).

171. Implication of central central nervous systems barriers impairment in amyotrophic lateral sclerosis patients : a sex-related difference

Hugo Alarcan(*)(1,2), Patrick Vourc'h (1,2), Lise Berton (1), Isabelle Benz-de Bretagne (1), Eric Piver (1), Christian Andres (1,2), Philippe Corcia (2,3), Charlotte Veyrat-Durebex (1,2), Hélène Blasco (1,2)

172. Investigating the suitability of urinary biochemical analytes to assess nutritional state in MND: a pilot study

Sarah Roscoe* (1), Elaine Kabucho Kibirige (1), Alexandra Grove (1), Scott Allen (1), Christopher McDermott (1), Theocharis Stavroulakis (1).

173. Lys-acetylated PPIA as a possible translational biomarker of ALS

Stefano Fabrizio Columbro* (1), Serena Scozzari (1), Laura Pasetto (1), Giovanni De Marco (2), Alfredo Cagnotto (3), Alessia Lanno (4), Alice Passoni (4), Renzo Bagnati (4), Mario Salmona (3), Andrea Calvo (2), Valentina Bonetto (1)

174. Macrophage inclusions in cerebrospinal fluid following treatment initiation with antisense oligonucleotide therapies in motor neuron diseases

Maximilian Vidovic (1)*, Mario Menschikowski (2), René Günther (1,3)

175. Multiparametric MRI texture and diffusion tensor imaging of the corpus callosum and cortico-spinal tract as a potential biomarker for PLS

Simon Witzel* (1), Hans-Peter Müller (1), Veronika Micca (1), Johannes Dorst (1), Joachim Schuster (1), André Huss (1), Hayrettin Tumani (1), Jan Kassubek (1), Albert C Ludolph (1)

176. Plasma Neurofilament light chain levels associate with nutritional status and disease progression in Amyotrophic Lateral Sclerosis

Aida Zulueta, Veronica Pischetola, Giuliana Federico, Jacopo Lanzone, Rachele Piras, Debora Pain, Paola Mariani, Aurora Ferrante, Elena Gattico, Eugenio Agostino Parati, Christian Lunetta (*)

177. Precision Medicine for Amyotrophic Lateral Sclerosis: Combinatorial Analysis of Patient Genomes

A. Malinowski1, K. Taylor1, C. Stubberfield1

178. Predictors of SOD1-ALS progression: role of sex and cancer from two population-based registries

Giulia Gianferrari (3), Ilaria Martinelli (1,2), Jessica Mandrioli (3,1), Elisabetta Zucchi (3,1), Andrea Ghezzi (3), Cristina Moglia (4), Umberto Manera (4), Luca Solero (4), Rosario Vasta (4), Antonio Canosa (4,5), Maurizio Grassano (4,5), Maura Brunetti (5), Letizia Mazzini (6), Fabiola De Marchi (6), Cecilia Simonini (1), Nicola Fini (1), Marco Vinceti (3,7,8), Adriano Chiò (4,5), Andrea Calvo (4,5)

179. Prognostic prediction models in motor neuron disease: a systematic review

Harry McDonough* (1), Lizzie Coates (1), Haris Stavroulakis (1), Dennis Wang (1), Christopher McDermott (1)

180. Prognostic Value of Serum Troponin T Elevations in a real life Cohort of ALS Patients

Rachel Fabian*(1), Sarah Bernsen(1), Saman Barakat(1), Teresa Koch(1), Barbara Radermacher(1), Patrick Weydt(1,2)

181. Relationships Between Brain Iron and Blood Markers for Iron and Inflammatory Status in Cognitively Healthy Adults.

Holly Spence * (1), Stephanie Mengoa-Fleming (1), Alan Sneddon (2), Christopher J. McNeil (1), and Gordon D. Waiter (1)

182. SerpinA1 as prognostic biomarker in Amyotrophic Lateral Sclerosis: an exploratory study

Andrea Ghezzi* (1), Ilaria Martinelli (2,3), Elisabetta Zucchi (2,4), Cecilia Simonini (2), Giulia Gianferrari (4), Roberta Bedin (2,5), , Nicola Fini (2), Serena Carra (1,5), Jessica Mandrioli (1,2,5)

183. T cell responses at diagnosis of amyotrophic lateral sclerosis predict disease progression

Solmaz Yazdani*(1), Christina Seitz(1), Can Cui(1), Anikó Lovik(1), Lu Pan(2), Fredrik Piehl(3), Yudi Pawitan(2), Ulf Kläppe(3), Rayomand Press(3), Kristin Samuelsson(3), Li Yin(2), Trung Nghia Vu(2), Anne-Laure Joly(3), Lisa S. Westerberg(4), Björn Evertsson(3), Caroline Ingre(3), John Andersson(1) & Fang Fang(1)

184. Thoracic spinal cord gray matter atrophy as a sensitive marker for early ALS

Maria Janina Wendebourg (* 1,2,7), Matthias Weigel (3,4,1,2,7), Claudia Weidensteiner (3,4), Laura Sander (1,2,7), Eva Kesenheimer (1,2,7), Nicole Naumann (1), Tanja Haas (3), Philipp Madoerin (3), Nathalie Braun (5), Christoph Neuwirth (5), Markus Weber (5), Kathleen Jahn (6), Ludwig Kappos (2,7), Cristina Granziera (1,2,7), Kathi Schweikert (1), Michael Sinnreich (1,8), Oliver Bieri (3,4), Regina Schlaeger (1,2,7)

185. Tongue strength as a biomarker of bulbar dysfunction and progression in ALS patients

Lidia, Bojtos*(1)Sílvia, Susín Calle(1)Julia, Peris Subiza(1)Bernat, Bertran Recasens(1)Anna, Guillén-Solà(2)(3)Juana María, Martínez Llorens(4)(5)Ana, Balaña Corberó(4)Montserrat, Villatoro Moreno(6)Greta, Garcia Escobar(1)Miguel, Angel Rubio(1)(7)

186. Transcranial Magnetic Stimulation (TMS) as a potential early biomarker of Amyotrophic Lateral Sclerosis (ALS): a 12-month longitudinal clinical study

Anna Carobin*(1), James Bashford(1), Viviana Santoro(1), Harry Clark(1), Lorenzo Rocchi(1), Isabella Premoli(1), Charles Large(2), Mark Richardson(1), Chris Shaw(1);

187. Treatment with Tofersen of a patient carrying a heterozygous novel frameshift SOD1 mutation (p.Ser108LeufsTer15) causing a premature termination codon

Aida Zulueta*, Veronica Pischetola*, Giuliana Federico, Jacopo Lanzone, Rachele Piras, Eugenio Agostino Parati, Christian Lunetta (*)

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188. Widespread alterations in Fast Amyotrophic Lateral Sclerosis Progressors: A Brain DTI and Sodium MRI Study

Mohamed Mounir El Mendili* (1,2), Aude-Marie Grapperon (2,3), Rémi Dintrich (1,2,3), Jan Patrick Stellmann (1,2), Jean-Philippe Ranjeva (1,2), Maxime Guye (1,2), Annie Verschueren (3), Shahram Attarian (3), Wafaa Zaaraoui (1,2).

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189. A Case of Juvenile Amyotrophic Lateral Sclerosis Carrying variants in TARDBP and NEK1: A peculiar phenotype and discussion of oligogenic model in ALS

Daniel Sánchez-Tejerina * (1,2,3), Juan Luis Restrepo-Vera (1,2), Eulalia Rovira-Moreno (4), Marta Codina-Sola (4), Verónica López-Diego (1,2), Arnau Llauradó (1,2), Javier Sotoca (1,2), María Salvadó (1,2), Núria Raguer (5), Elena García-Arumí (2), Raúl Juntas-Morales (1,2,3).

190. A multicenter prospective observational study to develop low-burden high-frequency prognostic digital speech biomarkers in ALS and FTD – PROSA

Johannes Tröger1, Judith Baltes3, Ebru Baykara1, Elisabeth Kasper5,7, Martha Kring5, Nicklas Linz1, Jessica Robin2, Simona Schäfer1, Anja Schneider3,8, Andreas Hermann4,5,

191. A MULTI-CENTER, RANDOMIZED, DOUBLE-BLIND, PLACEBO-CONTROLLED MULTIPLE ASCENDING DOSE STUDY TO EVALUATE THE SAFETY AND TOLERABILITY OF QRL-201 in ALS

Angela Genge(1)*, Kristiana Salmon(1), John Polzer(1), Clárida Martínez(1), Bryan Boggs(1), Victoria Eon(1), Rakhee Ganti(1), Sandy Hinckley(1), Kristina Johnson(1), Daniel Elbaum(1)

192. A MULTI-OMICS APPROACH TO STUDY MONOZYGOTIC TWINS DISCORDANT FOR AMYOTROPHIC LATERAL SCLEROSIS

Martina Tosi*(1), Miriam Zuccalà (1), Francesco Favero (1), Lucia Corrado (1), Roberta Croce (1), Chiara Basagni (1), Nadia Barizzone (1), Laura Follia (1), Andrea Saul Costa (1), Fabiola De Marchi (2), Elena Chinni (3), Letizia Mazzini (2), Davide Corà (1), Maurizio Leone (3), Sandra D'Alfonso (1)

193. A novel GRN mutation in an Italian patient with non-fluent variant of primary progressive aphasia at onset: a longitudinal case report

Veronica Castelnovo*,(1) Elisa Canu,(1) Laura Pozzi,(2) Francesca Vignaroli,(8) Edoardo Gioele Spinelli,(1,3) Teuta Domi,(2) Giacomo Tondo,(9) Cristoforo Comi,(9) Paola Carrera,(6) Nilo Riva,(2,4) Massimo Filippi,(1,3-5,7) Federica Agosta(1,3,7)

194. A Novel Machine-Learning Based Clinical Trial Subgroup Analysis Tool

Danielle Beaulieu, Kelly Smith, and David L. Ennist

195. A Phase 2 Trial of Pridopidine in ALS

Jeremy Shefner * (1), Bjorn Oskarsson (2), Melanie Quintana (3) , Ben Saville(3), Eric Macklin (4), Lori Chibnik (4), Michal Geva (5), Michael Hayden (5), James Berry (4), Sabrina Paganoni (4), Merit Cudkowicz (4) on behalf of the HEALEY ALS Platform Trial Study Group.

196. A randomized, double-blind, placebo-controlled, phase 2 study to assess safety, tolerability, and efficacy of RT001 in patients with ALS

Daphne Weemering* (1), Andrew Dannenberg (2), Anil Kumar (2), Mark Midei (2), Mikhail Shchepinov (2), Peter Milner (2), Vidhya Gopalakrishnan (2), Leonard van den Berg (1), Ruben van Eijk (1,3)

197. A weight loss risk prediction model in ALS

David G. Lester*(1), Martin R. Turner(1), Kevin Talbot(1), Alexander G. Thompson(1)

198. Acceptability of a Wearable Device to Evaluate Progression in People with Motor Neuron Disease

Emily Beswick* (1,2,3), Alexander Christides (1,2,3), Alexander Symonds (1,2,3), Michaela Johnson (1,2,3), Thomas Fawcett (4), Judith Newton (1,2,3), Dawn Lyle (1,2,3), Christine Weaver (1,2,3), Siddharthan Chandran (1,2,3,5) & Suvankar Pal (1,2,3).

199. Acces barriers for dignified death through euthanasia in Colombia

Nathalia Pérez-Becerra (1), Camilo Mendoza-Pulido (1,2), Martha Peña (2)*

200. Adolescents' need for professional support when living with a parent with ALS – based on both the adolescents' and the parents' experiences

Nina Malmström* (1), Birgitta Jakobsson Larsson (2, 3), Stefan Nilsson (1, 4), Joakim Öhlén (1,4, 5), Ingela Nygren (3, 6), Peter M. Andersen (7) & Anneli Ozanne (1, 8)

201. ADULT-ONSET OF FUS-RELATED AMYOTROPHIC LATERAL SCLEROSIS PATIENTS: PHENOTYPE AND GENOTYPE CHARACTERIZATION

Giovanni Colacicco* (1), Andrea Lizio (1), Lorena Mosca (2), Francesca Gerardi (1), Valeria Sansone (1), Federica Cerri (1)

202. Age and gender distribution in three aggressiveness strata in 653 Thuringian ALS patients

Alica Terörde (*) (1), Carolina Gerke (1), Meret Herdick (1), Linus Falge (1), Robert Steinbach (2), Anne-Kathrin Rödiger (2), Prof. Julian Großkreutz (1)

203. ALS and the Gut-Brain Axis: A systematic review and meta-analysis assessing the relationship between amyotrophic lateral sclerosis, the gut and its mi

Fergal M. Waldron (1*), Alexander T Jackson (1), Rollo G. Press (1), Jenna M. Gregory (1).

204. ALSFRS-R-SE: an adapted, annotated, and self-explanatory version of the revised amyotrophic lateral sclerosis functional rating scale

André Maier* (1), Matthias Boentert (2, 3), Peter Reilich (4), Simon Witzel (5), Susanne Petri (6), Julian Großkreutz (7), Moritz Metelmann (8), Paul Lingor (9), Isabell Cordts (9), Johannes Dorst (5), Daniel Zeller (10), René Günther (11, 12), Tim Hagenacker (13), Torsten Grehl (14), Susanne Spittel (15), Joachim Schuster (5, 16), Albert Ludolph (5, 16) & Thomas Meyer (1, 15) for the MND-NET consensus group

205. Alterations of spontaneous speech in primary progressive aphasia variants: a neuropsychological and brain MRI study

Veronica Castelnovo*,(1) Elisa Canu,(1) Sofia Santicioli,(1) Elena Gatti,(1) Alessandra Lamanuzzi,(1) Silvia Basaia,(1) Maria Antonietta Magno,(1) Giuseppe Magnani,(2) Francesca Caso,(2) Paola Caroppo,(6) Sara Prioni,(6) Cristina Villa,(6) Stefano Francesco Cappa,(7,8) Federica Agosta,(1,2,5) Massimo Filippi(1-5)

206. Amyotrophic Lateral Sclerosis and Lead

Halil Atilla İdrisoğlu(*)(1), Şeyda Sarıgöl(1), Hayim Babul(1), Elif Yıldırım(2)

207. An observational cohort study on pulmonary function in adult patients with 5q-spinal muscular atrophy under nusinersen therapy

Bogdan Bjelica (1) *, Camilla Wohnrade (1), Alma Osmanovic (1,), Olivia Schreiber-Katz (1), Susanne Petri (1)

208. Analytical Validation of Siemens Healthineers Atellica® IM NfL Assay and its Clinical Utility in People with SOD1-ALS in the VALOR Study (PREF POSTER)

Dan Bartlett* (1), Kristina H. Holmberg (1), Aileen Shields (2), Stephen Lee (3), Manjit McNeill (4), Peng Sun (1), Stephanie Fradette (1), Malcolm Shields (2), Danielle Graham (1)

209. Analyzing the use of statins and survival in people living ALS using real-world data

Alex Berger*1, Matteo Locatelli1, Christian Lunetta2, Alexander Sherman1,3

210. Arable land and vineyard proximity influenced ALS risk and age at onset

Umberto Manera(1,2), Stefano Callegaro(1), Rosario Vasta(1), Maurizio Grassano(1), Antonio Canosa(1,2,3), Fabiola De Marchi(4), Letizia Mazzini(4), Cristina Moglia(1,2), Adriano Chiò(1,2,3), Andrea Calvo(1,2)

211. Associations between cognitive performance and glucose metabolism in ALS patients

Annaliis Lehto(1,2*), Julia Schumacher(2,3), Elisabeth Kasper(2,3), Stefan Teipel(2,4), Andreas Hermann(1,2), Jens Kurth(5), Bernd Joachim Krause(5), Johannes Prudlo(2,3)

212. Autonomic Involvement in Spinal Muscular Atrophy: An exploratory study

Arman Çakar*, Hacer Durmuş, Yeşim Parman

213. Availability of ALS medical care in Croatia and perception of the future treatments

Mirea Hancevic* (1), Barbara Sitas (1), Hrvoje Bilic (1), Ervina Bilic (1)

214. Brain metabolism in symptomatic and asymptomatic C9orf72 repeat expansion carriers: a cross-sectional 1H and 31P magnetic resonance spectroscopy study

Henk-Jan Westeneng(1)*, Bram Nitert(1), Kevin van Veenhuijzen(1), Carrie Wismans(1), Graziella Donatelli(2), Harold Tan(1), Peter Luijten(3), Dennis Klomp(3), Alex Bhogal(3), Jan Veldink(1), Jannie Wijnen(3), Leonard van den Berg(1)

215. C9orf72 in the Spanish ALS-FTD spectrum

Daniel Borrego-Hernández (*)(1), Laura Expósito-Blázquez (1), Oriol Dols-Icardo (2) Celia Romero-Haro (1), Lucía Galán-Dávila (3), Sara Llamas-Velasco (4), Marta González-Sánchez (4), Alberto Villarejo-Galende (4), José Luis Muñoz-Blanco (5), David A. Pérez-Martínez (1, 4), Miguel Ángel Martín-Casanueva (6), Jesús Esteban-Pérez (1) and Alberto García-Redondo (1)

216. CARE-MND – A highly curated national integrated care, audit, and research platform for MND

*Isaac Chau(1),(2),(3), Jessica Gill(1),(2),(3), Judith Newton(1),(2),(3), Colin Smith(1),(2),(3), Siddharthan Chandran(1),(2),(3), Suvankar Pal(1),(2),(3), CARE-MND Consortium(4)

217. Characterization of FTLD spectrum through advanced diffusion-weighted MRI metrics: a connectome approach

Silvia Basaia*,(1) Federica Facente,(1) Stefano Pisano,(1,11) Camilla Cividini,(1) Edoardo Gioele Spinelli,(1,4) Elisa Canu,(1) Veronica Castelnovo,(1) Alma Ghirelli,(1,5) Giuseppe Magnani, (2) Francesca Caso,(2) Nilo Riva,(2,4,5) Paola Caroppo,(7) Sara Prioni,(7) Cristina Villa,(7) Lucio Tremolizzo,(8) Ildebrando Appollonio,(8) Federico Verde,(9) Vincenzo Silani V,(9,10) Massimo Filippi,(1-4,6) Federica Agosta(1,2,6)

218. Characterization of the Enrolled Population in the Phase 3 PHOENIX Trial in Amyotrophic Lateral Sclerosis: Preliminary Results

Leonard van den Berg* (1), Lahar Mehta (2), Ryan Miller (2), Suzanne Bijl (3), Feifan Zhang (2), Sabrina Paganoni (4,5)

219. Clinical and prognostic characterisation of patients with amyotrophic lateral sclerosis in Austria – a single-centre cohort study

Omar Keritam1,2,*, Haluk Caliskan1,2, Sigrid Klotz1,2, Merve Sener1,2, Fiona Jäger1,2, Rosa Weng1,2, Chiara Paternostro1,3, Ellen Gelpi1,2, Martin Krenn1,2, Gudrun Zulehner1,2, Jakob Rath1,2, Fritz Zimprich1,2, Hakan Cetin1,2

220. Clinical description of C9Orf72-mutated ALS patients followed in a large and representative Center for Motor Neuron Diseases of central Italy

Francesca Bianchi (*) (1), Lucrezia Becattini(1) ,Lorenzo Fontanelli L(1), Caterina Meoni(1)., Antonella Fogli (2), Gabriele Siciliano(1)

221. CNM-Au8® Preserved ALS Patient Function and Slowed Disease Progression in the OLE of the Phase-2 RESCUE-ALS Trial: New Data Through to 120 Weeks

Steve Vucic PhD, DSc, FRACP, FAHMS (1), Parvathi Menon PhD, FRACP (1), William Huynh PhD, FRACP (2), Colin Mahoney, PhD, MB, MRCPI (2), Karen S. Ho, PhD MSc (3), Marjan Sepassi, PharmD (*3), Austin Rynders, RN (3), Alan Hartford, PhD (3), Robert Glanzman, MD FAAN (3), Michael T. Hotchkin (3), Matthew C. Kiernan PhD, DSc, MBBS, FRACP, FAHMS (2).

222. Co-creation of new digital outcome measures in amyotrophic lateral sclerosis

*Navdeep Sahota(1), Elisa Ferrer-Mallol(1), Dirk De Valck(2), Evy Reviers(2), Elin Haf Davies(1)

223. Coexistence of Amyotrophic Lateral Sclerosis and autoimmune diseases: two cases

Lucrezia Becattini (*1), Francesca Bianchi (1), Lorenzo Fontanelli (1), Beatrice Giovannini (1), Caterina Meoni (1), Gabriele Siciliano (1) .

224. Coexistence of Amyotrophic Lateral Sclerosis and Pseudoxanthoma Elasticum in two siblings

Caterina Meoni (*)(1), Francesca Bianchi (1), Lucrezia Becattini (1), Lorenzo Fontanelli (1), Beatrice Giovannini (1), Gabriele Siciliano (1)

225. Colchicine treatment in Amyotrophic Lateral Sclerosis: a phase 2 multicenter, randomized, controlled, double-blind clinical trial

Jessica Mandrioli (1,2), Roberto D'Amico (3,4), Valeria Crippa (5), Serena Carra (1), Valentina Bonetto (6), Orietta Pansarasa (7), Cristina Cereda (8), Giulia Gianferrari (1,2), Elisabetta Zucchi (2,9), Riccardo Cuoghi Costantini (3), Ilaria Martinelli (2,10), Cecilia Simonini (2), Roberto Vicini (3), Francesca Trojsi (11), Nicola Ticozzi (12), Luca Diamanti (13), Mario Sabatelli (14,15), Eleonora Dalla Bella (16), Isabella Laura Simone (17), Eveljn Scarian (7), Laura Pasetto (6), Alberto Doretti (12), Co-ALS investigators group, Angelo Poletti (5).

226. Combining patient preferences, function and survival in clinical trials for amyotrophic lateral sclerosis

Ruben P.A. van Eijk*, Leonard H. van den Berg, Ying Lu

227. Correlation of cardiac Troponin T with parameters of body plethysmography, polysomnography and diaphragmatic ultrasound in patients with Amyotrophic L

Teresa Koch* (1), Sarah Bernsen (1,4), Saman Mohamed Ali Barakat (1), Leonie Weinhold (2), Franz-Wilhelm Koch (3), Patrick Weydt (1)

228. Course of cognitive and behavioral impairment, quality of life and caregiver status in Amyotrophic Lateral Sclerosis (ALS): A 6-Month Follow-Up Study

Greta García-Escobar(1,2), Bernat Bertran-Recasens (1), Sandra Blavi-Pujol (3), Anna Guillen- Sola (4,5), Juana María Martínez Llorens (6,7), Ana Balaña Corberó (6), Montserrat Villatoro Moreno (8) and Miguel Ángel Rubio (1,9)

229. Crushing riluzole tablet: methodology of a home-simulation experiment to evaluate the loss of powder and active principle in this product manipulation

Antonella Casiraghi (1), Adriano Chiò (2)*

230. Decision making about gastrostomy placement in MND: a survey of UK healthcare professionals beliefs and practice

Sean White (1)*, Professor Alicia O'Cathain (2), Dr Vanessa Halliday (2), Mike Bradburn (2), Professor Christopher J McDermott (1)

231. Decision-making about gastrostomy placement and ventilation in motor neuron disease (MND) care: a qualitative evidence synthesis

Sean White (1)*, Professor Alicia O'Cathain (2), Dr Vanessa Halliday (2), Dr Liz Croot (2), Professor Christopher J McDermott (1)

232. DESIGN OF AN INTERNATIONAL, PHASE 3 OLE STUDY TO INVESTIGATE THE LONG-TERM SAFETY OF DAILY ORAL EDARAVONE (FNP-122) IN ALS: THE ADOREXT STUDY

Leonard van den Berg (1), Ruben Van Eijk (2), Carla Varona (3), Esther Miralles (4), Tania Nadal (5), Cristina Tarragó (5), Nuria Albareda (6)

233. Designing a neurophysiological module for multicenter early phase clinical trials in people with ALS

Boudewijn T.H.M. Sleutjes (1), H. Stephan Goedee (1), Romy L. Verschoor (1), Tommy M. Bunte (1), Ruben P.A. van Eijk (1), Leonard H. van den Berg (1)

234. Developing a Remote stimulation-free Motor Unit Number Estimate (REMUNE).

Judith Bilgorai(*), James Bashford (1), Anne Vanhoestenberghe (1), Gerrard Rafferty (1), Maoqi Chen (2), Ping Zhou (2), Yuebo Song (1), Christopher Shaw (1)

235. Developing and evaluating complex digital interventions for people with MND: A case study of the Telehealth in MND service.

*Liam Knox (1), Deirdre Murray (2), Deirdre Gilsenan (2), Miriam Galvin (2), David Murphy (2), Orla Hardiman (2), Esther Hobson (1), and Chris McDermott (1).

236. Diagnosis and symptom characteristics of diverse patient populations with amyotrophic lateral sclerosis in the USA

Olga Sánchez-Soliño(1)*, Joseph Guilfoyle(1), Yunhua Fan (1), Joey Boiser (1), Lisa Vinikoor-Imler (1)

237. Digital outcome measure assessing the motor function of patients with amyotrophic lateral sclerosis

Y. Bechichi (1), M. Poleur (2), G. Parinello (1), L. Buscemi1, S. Delstanche (3), A. Maertens de Noordhout (3), O. Bouquiaux (3), I. Lievens (3), D. Eggenpieler (1), L. Servais (2, 4)

238. Directly measuring network function during social cognition in ALS using EEG during the Reading the Mind in the Eyes Task

Roisin McMackin*(1)(2), Serena Plaitano(2), Marjorie Metzger(2), Vlad Sirenko(2), Rosie Giglia(2), Prabhav Mehra(2), Yasmine Tadjine(2), Saroj Bista(2), Mark Sabbagh(3), Niall Pender(2), Bahman Nasseroleslami(2), Orla Hardiman(2)

239. Double trouble: clinical and neuroimaging features in a case of frontotemporal dementia with C9orf72 expansion and progranulin mutation

240. Double-blind randomised study to assess IFB-088 plus riluzole vs placebo plus riluzole in patients with bulbar-onset amyotrophic lateral sclerosis

241. Effect of edaravone therapy in Turkish amyotrophic lateral sclerosis (ALS) patients

Halil Atilla İdrisoğlu(*)(1), Şeyda Sarıgöl(1), Elif Yıldırım(2), Reşit Çelik(2), Şenay Vural Korkut(2), Aslıhan Günel(2)

242. Effect of tauroursodehoxycholic acid on survival and safety in amyotrophic lateral sclerosis: a retrospective population-based cohort study

Elisabetta Zucchi(1,2)*, Umberto Maria Musazzi(3), Guido Fedele(4), Ilaria Martinelli(1,5), Giulia Gianferrari(1,6), Nicola Fini(1), Andrea Ghezzi(1,6), Maria Caputo(1,6), Elisabetta Sette(7), Veria Vacchiano(8), Lucia Zinno(9), Elena Canali(10), Marco Vinceti(11), Salvatore Ferro(12), Jessica Mandrioli(1,6)

243. Enlarged perivascular spaces in the midbrain are more prevalent in ALS and are associated with primary motor cortex atrophy

Bryan Connolly (1,2)*, Roberto Duarte Coello (3), Charlotte Zejlon (1,2), Stefan Sennfält (2,4), Russell Ouellette (1,2), Benjamin Victor Ineichen (2), Georg Walther (2), Sven Petersson (1,6), Sebastian Lewandowski (2), Joanna Wardlaw (3), Johannes Finnsson (1,2), Caroline Ingre (2,4), Tobias Granberg (1,2).

244. Epidemiological and Phenotypic features in the French ALS population: A prospective study on a cohort of 1000 index cases.

P Corcia1, J Cassereau2, G le Masson3, M LefIlliatre4, N Guy5, A Jacquin-Piques6, V Danel7, P Couratier8, G Lautrette8, E Bernard9, A Vershueren10, W Camu11, F Esselin11, E de La Cruz11, S Pittion12, MH Soriani13, M del Mar Amador14, JP Camdessanché15, MC Fleury16, P Cintas17, A Choumert18, P Sauleau19, S Genestet20, K Mouzat21, AL Fauret22, P Vourc'h23.

245. Epidemiology and Patient characteristics of Amyotrophic Lateral Sclerosis (ALS) in France using the SNDS database

Katie Stenson *(1), Varant Kupelian (1), Hicham Hadjrabia (2), Agathe Doutriaux (2), Daphné Avot (2), Seham ISSA (3), Sophie Marguet (3), Frederic Bernard (3), Enkhgerel Nasanbat (3), Gérard de Pouvourville (4), Philippe Couratier (5), Philippe Corcia (6)

246. Epidemiology of Serum Neurofilaments in Amyotrophic Lateral Sclerosis – Population-based Evidence For Diagnostic and Prognostic Utility

Simon Witzel* (1), André Huss (1), Dietrich Rothenbacher (2), Gabriele Nagel (2), Johannes Dorst (1), Joachim Schuster (1), Ulrike Weiland (1), Raphael Peters (2), Dorothée Lulé (1), Hayrettin Tumani (1), Angela Rosenbohm (1), Albert Ludolph (1), ALS Registry Swabia Study Group

247. Evaluating single and multiple ascending doses of WVE-004 in C9orf72-associated ALS and FTD: results from the FOCUS-C9 Trial

Michael Tillinger1, Merit Cudkowicz2, Jonathan D. Rohrer3, Leonard H. van den Berg4, Johnathan Cooper-Knock5, Simon Ducharme6, Kenechi Ejebe7, Andrew Hart1, Yili Pritchett1, Xiao Shelley Hu1, Padma Narayanan1, Maneet Singh1, Stephen L. Lake8, Michael Panzara9, Anne-Marie Li-Kwai-Cheung1

248. Evaluating the Efficacy and Safety of Tofersen in Adults with ALS and a SOD1 Mutation: Results from the Phase 3 VALOR Trial and Open-Label Extension

Philip Van Damme* (1),Timothy M. Miller (2), Merit Cudkowicz (3), Pamela J. Shaw (4), Angela Genge (5), Gen Sobue (6), Ivan Nestorov (7), Laura Fanning (7), Peng Sun (7), Luan Lin (7), Manjit McNeill (8), Danielle Graham (7), Stephanie Fradette (7), on behalf of The VALOR and OLE Working Group

249. Evaluating the Safety, Tolerability, and Pharmacokinetics of QRL-101 in Two Phase 1 Studies: QRL-101-01 in Healthy Adults and QRL-101-02 in Adults With ALS

Angela Genge1, Bryan Boggs1, Rakhee Ganti1, Sandy Hinckley1, Kristina Johnson1, Hannah Kaneb1, John Polzer1, Samantha Rubino1, Kristiana Salmon1, and Daniel Elbaum1

250. Evidence for a Survival Benefit in ALS with CNM-Au8 Treatment: Updated Results from the RESCUE-ALS Trial Long-Term Open Label Extension

Steve Vucic PhD, DSc, FRACP, FAHMS (1), Parvathi Menon PhD, FRACP (1), William Huynh PhD, FRACP (2), Colin Mahoney, PhD, MB, MRCPI (2), Karen S. Ho, PhD MSc (3), Marjan Sepassi, PharmD (3), Austin Rynders, RN (3), Alan Hartford, PhD (3), Robert Glanzman, MD FAAN (3), Michael T. Hotchkin (3), Matthew C. Kiernan PhD, DSc, MBBS, FRACP, FAHMS (2)

251. Examination of Age, Period and Cohort Effects using a Partial Least Squares Regression (PLSR) model

Robert McFarlane(1)*, Mark Heverin(1), Cathal Walsh(1), Orla Hardiman(1)

252. EXAMINING COGNITION AND BEHAVIOURAL PROFILE IN ALS-FTD SPECTRUM

María José Gil Moreno (1)*, Juan Ignacio López Carbonero (1,2), María Valles Salgado (1), Angélica Larrad Sainz (3), Juan Miguel Godoy Corchuelo (2), Alejandro Horga Hernández (1), Lucía Galán Dávila (1), Antonio Guerrero Sola (1), Javier Olazarán Rodríguez (4), Miriam Eimil Ortiz (5), Pilar Alcántara Miranda (5), Jorge Matías-Guiu Guía (1), Jordi Matías-Guiu Antem (1), Silvia Corrochano Sánchez (2)

253. Exploring treatment burden and adherence to treatment in ALS patients: a prospective multicentric study

Andrea Lizio (1), Vania Campanella (1), Michela Nani, Veronica Pischetola (3), Valeria Sansone (1), Christian Lunetta (3), Luca Diamanti (2), Federica Cerri* (1)

254. Factors Impacting Trial Participation in People with Motor Neuron Disease

Emily Beswick*(1,2,3), Kay Johnson (1,2,3), Judith Newton (1,2,3), Rachel Dakin (1,2,3), Amy Stenson (1,2,3), Sharon Abrahams (3,4), Alan Carson 1,2, Siddharthan Chandran (1,2,3,5) & Suvankar Pal (1,2,3).

255. Familial ALS with a Novel ANXA 11- Mutation Presenting with a Proximal Pattern

Andrea Chici*, Markus Weber

256. First-in-human study of safety, tolerability, PK, and PD of SAR443820, a centrally penetrant RIPK1 inhibitor in healthy participants

Amy Eastenson (1), Agnes Hincelin-Mery (2), Pascale Lewanczyk (3), Cathy Cantalloube (2), Xavier Nicolas (4), Myriam Benamor (2), Robert Pomponio (5), Emmanuel Krupka (4), Dimitry Ofengeim (5), Li Xiong* (5), Nazem Atassi (5)

257. Gastrostomy tube – is it for me?' A web-based decision aid to support people with motor neurone disease considering a gastrostomy tube

Sally Wheelwright (1), Rose Maunsell (2), Sophia Taylor (2), Neil Drinkwater (3), Clare Erridge (4), Claire Foster (2), Maggi Hardcastle (5), Anne Hogden (6), Ian Lawson (2), Dominika Lisiecka (7), Christopher McDermott (8), Karen Morrison (9), Cath Muir (2), Alejandra Recio-Saucedo (2) and *Sean White (8).

258. Genetic characterization of primary lateral sclerosis

Eva M.J. de Boer*(1), Balint S. de Vries(1),Maartje Pennings(2), Erik-Jan Kamsteeg(2), Jan H. Veldink(1),Leonard H. van den Berg(1),Michael A. van Es(1)

259. Global Fundamental Rights in ALS/MND Survey: A look into the global results.

Cathy CUMMINGS(1), Jessica MABE(1)*

260. HTT and ATXN2 repeat expansions increase risk of ALS in a Norwegian ALS cohort: The GAIN study

Camilla Novy*(1), Øyvind Løvold Busk(1), Maria Fahlström(1), Karl Bjørnar Alstadhaug(2), Ingrid Kristine Bjørnå(3), Bahri Bunjaku(4), Natasha Demic(5), Heidi Øyen Flemmen(6), Ineke HogenEsch(7), Margitta T. Kampman(8), Grethe Kleveland(9), Helene Ballo Kvernmo(10,11), Sigve Strand Landa(1), Unn Ljøstad(12,13), Angelina Maniaol(14), Åse Hagen Morsund(15), Cathrine Goberg Olsen(1), Katrin Schlüter(16), Stephan Schuler(17), Ole-Bjørn Tysnes(18), Øystein Lunde Holla(1), Trygve Holmøy(19,20), Helle Høyer(1).

261. Hypothesis-free Mendelian Randomisation Identifies New Metabolites Linked to Risk of ALS

Elham Alhathli(1,3)*, Thomas H Julian(2), Calum Harvey(1), Project MinE sequencing consortium, Dennis Wang(4,5), Pamela J Shaw(1), Johnathan Cooper-Knock(1)

262. Impact of bulbar involvement on respiratory impairment in ALS patients

Sílvia Susín Calle* (1), Lidia Bojtos (1), Julia Peris Subiza (1), Bernat Bertran Recasens (1), Anna Guillen-Sola (2, 3), Juana María Martínez Llorens (4,5), Ana Balaña Corberó (4), Montserrat Villatoro Moreno (6), Greta Garcia Escobar (1), Miguel Angel Rubio (1,7)

263. Impact of COVID-19 pandemic on the progression and survival of patients with amyotrophic lateral sclerosis

Pilar H García-Casanova* (1), Pablo Pérez Martínez (2), Teresa Sevilla (2,3,4,5) Montserrat León (6), Rosalía Doménech (6), Juan F Vázquez-Costa (2,3,4,5).

264. Impairment of oculomotor functions in patients with early to advanced ALS

Elisa Aust* (1), Sven-Thomas Graupner (2), René Günther (1,3), Katharina Linse (1,3), Markus Joos (4), Julian Großkreutz (5), Johannes Prudlo (6,7), Sebastian Pannasch† (8), Andreas Hermann† (9,10)

265. Interdisciplinary approach in Amyotrophic Lateral Sclerosis: the case of the ALS Centre of Uruguay, the CE-LAU Program

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269. Light The Way: Development of an Online Support Platform for People at Risk of Genetic ALS/FTD in the UK and US

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273. Longitudinal remote monitoring of nocturnal heart-rate and respiration in people living with amyotrophic lateral sclerosis

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274. Long-term follow-up of five Turkish families with UBQLN2 mutations

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Thomas Marlow* (1), Raquel Martins (1), Katie Bowden (1), Pamela Shaw † (1), Laura Ferraiuolo † (1). † - These authors equally contributed to this work.

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280. Navigating through a changing sea: The emotional journey of current family carers of people living with MND

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281. Neurofilament light chain response during therapy with Tofersen in SOD1-related ALS – treatment experience in clinical practice.

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282. Neurological Soft Signs in unaffected relatives of people with familial ALS: an expanding endophenotype of neurological dysfunction in ALS kindreds

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283. Non-Invasive Mechanical Ventilation reduces the Motor Decline in Amyotrophic Lateral Sclerosis

Maurizio Grassano* (1); Emanuele Koumantakis (1,2);Umberto Manera (1,3); Antonio Canosa (1,3); Rosario Vasta (1); Palumbo Francesca (1); Giuseppe Fuda (1); Paolina Salamone (1); Giulia Marchese (1); Federico Casale (1); Lorena Charrier (2); Gabriele Mora (1); Cristina Moglia (1,3); Andrea Calvo (1,3); Adriano Chiò (1,3,4)

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285. Outreach to Spanish-speaking populations in Latin America to improve participation in clinical trials offered at the Clinical Research Unit (MNI)

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286. Patient and technical feasibility of real-world sampling of cognition and functional neurophysiology in ALS and FTD

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288. Patients' and caregivers' views of multidimensional and palliative care in amyotrophic lateral sclerosis – protocol of a German multicentre study

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289. People at Risk of Genetic ALS/FTD Want Pre-Symptomatic Treatment: Results of an Online Survey

Jean Swidler(1), Tucker Olson(1), Amy L Edelstein(1), Julie Granning(1), Cassandra Haddad(1), Mindy Uhrlaub(1), Paul Wicks(*1,2) on behalf of the "Genetic ALS & FTD: End The Legacy" Group

290. Peripheral immune signatures in ALS: a population-based study

Enrico Matteoni* (1), Maurizio Grassano (1), Umberto Manera (1, 2), Fabiola De Marchi (4), Margherita Daviddi (1), Luca Solero (3), Alessandro Bombaci (1), Francesca Palumbo (1), Rosario Vasta (1), Antonio Canosa (1, 2, 6), Paolina Salamone (1), Giuseppe Fuda (1), Federico Casale (1), Letizia Mazzini (4), Andrea Calvo (1, 2, 5), Cristina Moglia (1, 2), Adriano Chiò (1, 2, 5, 6)

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Jeffrey Rothstein, MD, PhD (1), Shari DeSilva, MD (2), Lorne Zinman, MD, MSc, FRCPC (3), Marvin Chum, MD, FRCPC (4), Adriano Chio, MD, FAAN (5), Albert C. Ludolph, MD (6), Gen Sobue, MD, PhD (7,8), Manabu Doyu, MD, PhD (9), Daniel Selness, RN, BA, MBA (10), Vesna Todorovic, MD, MPhil (11), Manabu Hirai, MS (12), Takahashi Fumihiro, PhD (12), Takeshi Sakata, MS (12), Art Wamil MD, PhD (10), Alejandro Salah, MD, PhD, MBA, MHA (13), Stephen Apple*, MD (13)

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Anna Mestre*(1), Raquel Horrillo (1), Estefania Alcaraz (1), Miquel Barceló (1), Laura Nuñez (1), Isabel Bravo (1), Montserrat Costa (1)

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294. Portable fixed dynamometry enables home-based, reliable assessment of muscle strength in patients with amyotrophic lateral sclerosis: a pilot study

Jordi W.J. van Unnik (1)*, Jaap N.E. Bakers (2), Steure Kokx (2), Leonard H. van den Berg (1), Johanna M.A. Visser-Meily (2), Anita Beelen (2), Ruben P.A. van Eijk (1)(3)

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Jiajing Hu* (1,2), Ammar Al-Chalabi (2,3), Alfredo Iacoangeli (1,2,4)

296. Prodromal phase of sporadic ALS: a patient and caregiver perspective

Lisa Kraneburg*(1), Iris van Wijk(1), Henk-Jan Westeneng(1), Ruben van Eijk (1), Jan Veldink (1), Leonard van den Berg(1)

297. Proximal Hirayama Disease

Andrea Fortuna* (1), Giulia Gianferrari (1,2), Alen Bebeti (1), Lorenzo Blasi (1), Francesca Calvi (1), Serenella Salmaso (1), Gianni Soraru(1)

298. Quantitative evaluation of factors contributing to the results of RNS Test on ALS patients at initial visit.

Jinghong Zhang*(1), Yahui Zhu(1), Fei Yang(1), Mao Li(1), Ying Zhang(2), Xusheng Huang(1)

299. Quinine for the treatments of muscle cramps in amyotrophic lateral sclerosis; a randomized, placebo controlled, double-blind cross-over trial

Christoph Neuwirth* (1), Ursula Schneider, (1) Markus Weber (1)

300. Remote monitoring of accelerometry for amyotrophic lateral sclerosis detects differences in disease progression and survival

Jordi W.J. van Unnik* (1), Daphne Weemering (1), Leonard H. van den Berg (1), Ruben P.A. van Eijk (1)(2)

301. Respiratory function conservation in ALS is potentially linked to modulation of the innate immune system

Michael McGrath(*)(1,2), Rongzhen Zhang (1), Ari Azhir (2), Paige Bracci (1), Bruce Forrest (2).

302. Results from first-in-human study of VRG50635, a PIKfyve inhibitor for treatment of ALS, in healthy adult volunteers

Lars Smits(1), Jules Heuberger(1), Philip Kremer(1), Sujata Sankar(2), Payal Nanavati(2), Bruce Morimoto(2), Joanna Haas (2), Alistair Stewart(2), Irene Choi(2), Robert H. Scannevin(2), Brad Wong(2), Brian Shook(2), Robert Galemmo(2), Diego Cadavid(2)*

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Danilo Tornabene* (1) , Giuseppe Fiamingo (1) , Marianna Collesi (2), Eleonora Rigoni (2) , Stella Gagliardi (2) , Elena Ballante (2), Orietta Pansarasa (2), Silvia Scaranzin (2) , Matteo Gastaldi (2), Luca Diamanti (2).

304. Role of traumatic injury in Amyotrophic Lateral Sclerosis: a "double-hit" pathogenic model of neurodegeneration

Karthik Baskar* (1), Sandeep Rajkumar (1), Prof. Tobias M. Böckers (1)(2), Dr. Alberto Catanese (1)(2)

305. Safety and Activity of Anti-CD14 Antibody IC14 (atibuclimab) in ALS: Experience with an Expanded Access Protocol

Dario Gelevski*, BS (1), Grace Addy, BS (1), Caroline Cohen, BS (1), Margot Rohrer, RN (1), Aimee Roderick, MPH, BSN RN (1), Erin Clampffer, RN (1), Allison Winter, BS (1), Judith Carey, RN (1), Jennifer Scalia, NP-BC (1), Megan Yerton, AB (1), Harli Weber, BS (1), Michael Doyle, BA (1), Neil Parikh, BA (1), Geli Kane, BA (1), Amy Ellrodt, PT (1), Katherine Burke, PT (1), Derek D'Agostino, BA (1), Ervin Sinani, BS (2), Hong Yu, MS (1), Alexander Sherman, MS (1), Jan Agosti, MD (2), Garry Redlich, BA LLB (2), Patrick Charmley, PhD (2), David Crowe, PhD (2), Mark Appleby, PhD (2), Brian Ziegelaar, PhD (2), Katherine Hanus, BS (3), Zhenhua Li, BS (3), Johnna Varghese, PhD (3), Suma Babu, MD (1), Katharine Nicholson, MD (1), Sarah Luppino, NP-BC (1), James Berry, MD MPH (1), Clare Baecher-Allan, PhD (3), Sabrina Paganoni, MD PhD (1, 4), Merit Cudkowicz, MD MSc (1)

306. Safety, Pharmacokinetics, and Pharmacodynamics of the Selective Glucocorticoid Receptor Modulator Dazucorilant in Healthy Volunteers

Kirsteen Donaldson (1), Joseph M. Custodio (2), Hazel J. Hunt (2)(*)

307. Sex-related differences in Amyotrophic Lateral Sclerosis: a brain 2-[18F]FDG-PET study

Umberto Manera(1,2), Alessio Martino(3), Alessandro Giuliani(4), Cristina Moglia(1,2), Rosario Vasta(1), Francesca Palumbo(1), Filippo De Mattei(1), Enrico Matteoni(1), Margherita Daviddi(1), Giuseppe Fuda(1), Giovanni De Marco(1), Marco Pagani(3,5), Andrea Calvo(1,2), Adriano Chiò(1), Antonio Canosa(1,2,3)

308. Social Cognition and Executive functions in Amyotrophic Lateral Sclerosis: a population-based study.

Francesca Palumbo*(1), Barbara Iazzolino(1), Laura Peotta(1), Antonio Canosa(1,2), Umberto Manera(1,2), Maurizio Grassano(1), Fuda Giuseppe(1), Casale Federico(1), Salamone Paolina(1), Vasta Rosario(1), Moglia Cristina(1,2), Adriano Chiò(1,2), Andrea Calvo(1,2).

309. SOD1-ALS: patient journey in the tofersen early access program

Maximilian Vidovic (1)*, Hanna Sophie Lapp (1), René Günther (1,2)

310. Stepwise functional brain architecture from disease epicenter correlates with atrophy in progressive supranuclear palsy

Edoardo Gioele Spinelli*,(1,2) Alma Ghirelli,(1,2,3) Ilaria Bottale,(3) Silvia Basaia,(1) Camilla Cividini,(1) Maria Antonietta Volontè,(2) Sebastiano Galantucci,(2) Giuseppe Magnani,(2) Francesca Caso,(2) Elisa Canu,(1) Veronica Castelnovo,(1) Paola Caroppo,(4) Sara Prioni,(4) Cristina Villa,(4) Keith A Josephs,(5) Jennifer L Whitwell,(6) Federica Agosta,(1,2,3) Massimo Filippi(1,2,3,7,8)

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Ram Miller (1), Marie-Emmanuelle Riviere (2) (*), Ozana Fischer (3), Douglas Hom (1), Francesca Callegari (2), Lucie Bruijn (4), Elisa Garcia-Garayoa (3), Ronenn Roubenoff (1), Wendy Su (5)

312. Systematic review of the methodological quality of clinical trials in Amyotrophic Lateral Sclerosis

Elisabetta Pupillo* (1), Serena Sassi (1), Emilio Arippol (1), Lorenzo Tinti (1), Eugenio Vitelli (1), Maurizio Leone (1), Elisa Bianchi (1)

313. Testing Spinal Motor Neuron Function Using Vibration-Evoked Persistent Inward Currents: A Preliminary Study

Matthew Mitchell (1) (*), Orla Hardiman (1) (2), Richard G Carson (3) Bahman Nasseroleslami (1)

314. The deltoid muscle – part of the monosynaptic pattern of pareses in ALS

Albert Ludolph, Veronika Klose, Johannes Dorst, Ulm Germany

315. The Edinburgh Brain Bank – a global leader in discovery science and reverse translation in motor neuron disease

Hatice Bozkurt(1)(2)(3), Judith Newton* (1)(2)(3) , Bhuvaneish Selvaraj(1)(2)(3), Suvankar Pal (1)(2)(3) , Siddharthan Chandran (1)(2)(3), CARE-MND Consortium (4), Colin Smith (1)(2)(3)(4)(5)

316. The Effect of the Fat Content of Food on the Pharmacokinetics and Safety of Utreloxastat Oral Solution Formulation in Healthy Volunteers

Shivakumar Patil* (1), Lan Gao (1), Diksha Kaushik (1), Mandar Dali (1), Aaron Tansy (1), Mayzie Johnston (1), and Ronald Kong (1)

317. The Impact of Non-Motor Symptoms in People with Motor Neuron Disease

Emily Beswick* (1,2,3), Deborah Forbes (1,2,3), Kay Johnson (1,2,3), Judith Newton (1,2,3), Rachel Dakin (1,2,3), Sharon Abrahams (3,4), Alan Carson (1,2), Siddharthan Chandran (1,2,3,5) & Suvankar Pal (1,2,3).

318. The Inclusion of Lytico-Bodig Biospecimens into the United States National ALS Biorepository

Paul Mehta MD(1)*, Lyle W. Ostrow, MD PhD(2), Moon Han PhD(1), Jaime Raymond MPH(1), Kathleen Wilsbach PhD(2), Kathryn Gallo(2), Theodore Larson MS(1), Ralph Garruto PhD(3) Kevin Horton DrPH(1)

319. THE INTEGRATED STRESS RESPONSE IS MODULATED BY EIF2B AGONIST DNL343: RESULTS FROM PHASE 1 HEALTHY SUBJECT AND PHASE 1B ALS PATIENT STUDIES

Linus D. Sun(1), Richard M. Tsai(1), Ernie Yulyaningsih(1), Melania H. Fanok(1), Maurits F.J.M. Vissers(2,3), Jules A.A.C. Heuberger(2), Brittany N. Flores(1), Fen Huang(1), Lesley Kane(1), Isaac V. Cohen(1), Shyeilla V. Dhuria(1), Meng Fang(1), Anthony A. Estrada(1), Maksim Osipov(1), Brent Willman-Yoswa(1), Romeo Maciuca(1), Sarah K. Dobbins(1), Roni Chau(1), Timothy K. Earr(1), Hoang N. Nguyen(1), Isabel A. Lopez(1), Danna Jennings(1), Kimberly Scearce-Levie(1), Tommy M. Bunte(4), Leonard H. Van Den Berg(4), Carole Ho(1), Geert Jan Groeneveld(2,3), Matthew D. Troyer(1)

320. The landscape of neurophysiological outcome measures in ALS interventional trials: a systematic review

Najma Ahmed (1), Mark Baker (2), James Bashford* (3)

321. The perception of unpleasant sensations of patients with amyotrophic lateral sclerosis during procedures required by clinical trials (PESALS)

Martina Sodano (*) (1), Alberto Doretti (1), Eleonora Colombo (1), Gianluca Demirtzidis (1), Francesco Gentile (1), Carla Amigoni (1), Francesca Tagliacarne (1), Laura Ingrande (2), Nicola Ticozzi (1), Vincenzo Silani (1), Debora Rosa (2).

322. The relationship between quality of life and cognitive or behavioural impairment in motor neuron disease: a systematic review

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323. The revised El Escorial Criteria (EEC) use in different countries

Nathalie Braun* (1), Ruben P A van Eijk (2), Leonard H van den Berg (2), Hardiman Orla (3), Philip Van Damme (4, 5), Adriano Chio (6,7), Markus Weber (1)

324. TIDALS : Protocol for a Randomized, Placebo-Controlled, Double-Blind Phase II Trial of Safety and Efficacy of the GSK-3 inhibitor Tideglusib in ALS

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325. Time from ALS symptom onset to key disease milestones for slow, intermediate, and fast progressors: a real-world cross-sectional survey

Paulos Gebrehiwet* (1), Johan Brekke (1), Stacy A. Rudnicki (1), Jennifer Mellor (2), Jack Wright (2), Lucy Earl (2), Nathan Ball (2), Halima Iqbal (2), Owen Thomas (2), Giorgio Castellano (2)

326. Tofersen in SOD1 ALS: the experience of Italian NeMO Centers.

Mario Sabatelli* (1), Federica Cerri (2), Riccardo Zuccarino (3), Agata Katia Patanella (1), Daniela Bernardo (1), Giulia Bisogni (1), Raffaella Tanel (3), Valeria Sansone (2), Massimiliano Filosto (4), Amelia Conte (1)

327. Tofersen Safety Experience From An Ongoing Global Expanded Access Program.

Sohail Malek (1)*, Alexandra Lovett (1), Thos Cochrane (1), Stephanie Fradette (1), Karen Smirnakis (1), Laura Fanning (1), Jennifer Inra (1)

328. Traditional respiratory measurement and the current state of the field

Conor Hayden (*) (1,2,4), Bruce Murphy (1,2,3), Orla Hardiman (4,5), Deirdre Murray (4,5)

329. Treatment with acetyl-l- carnitine: new prospective from an observational retrospective study

Serena Sassi* (1), Bianchi Elisa (1), Luca Diamanti (2), Maurizio Inghilleri (3), Elisabetta Pupillo (1) and the ALCAR study group.

330. Two novel SOD1 gene variants with aggressive ALS phenotype

Amelia Conte* (1), Serena Lattante (2,3), Giulia Bisogni (1), Daniela Bernardo (1), Agata Katia Patanella (1), Emiliana Meleo (1), Lucia Maria Porro (1), Marcella Zollino (2,3), Mario Sabatelli (1,4)

331. Untangling a diagnosis with overlapping phenotypes: Late Onset Tay-Sachs disease and two sporadic ALS cases in one family.

Magui Khazaal* (1), Nazli S Kara (1), Blanka Chylikova (1), Nela Navratilova (1), Daniel Baumgartner (2), Helena Jahnová (3), Helena Poupetova (3), Tomas Honzik (3), Radim Mazanec (2), Lenka Slachtova (1)

332. Validity and reliability measures of the Swedish version of the Edinburgh Cognitive and Behavioral ALS Screen (ECAS)

Juliette Foucher* (1,2), Anikó Lovik(3,4), Stefan Sennfält (2), Fang Fang (3), Caroline Ingre (1,2)

333. Volumetric Analysis of the Brainstem: Predictability of Respiratory and Bulbar Function in Amyotrophic Lateral Sclerosis

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334. Worldwide geographical distribution of clinical trials in amyotrophic lateral sclerosis based on information available in clinical trial databases

Beliu García-Parra * (1), Josep M. Guiu (2), Pilar Modamio (2), Eduardo L. Mariño (2), Mónica Povedano (3)

335. Yentl's syndrome, a real phenomenon in Amyotrophic Lateral Sclerosis (ALS)?

Tatiana Gómez Escobar* (1), Saioa Fernández Soberón (2), Alejandro Caravaca Puchades (3), Juan Francisco Vázquez Costa (4), Jesús Mora Pardina (5), Raúl Juntas Morales (6), Mónica Povedano Panadés (7)

336. Prevalence of Oropharyngeal Dysphagia and its perception in ALS patients.

S.A. Riera1,2, A. Prats1, V. Herrera1, E. Romero1,2, N.Virgili2,3, M. Povedano1,2

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337. Deletion of endothelial TDP-43 disrupts the vascular barrier triggering inflammation and hemorrhages in the central nervous system

Victor Arribas* (1), Yara Onetti (1), Marina Ramiro (2,3), Pilar Villacampa (1), Ofelia Martinez (2,3), Heike Beck (4), Markus Sperandio (4), Bettina Schmid (5), Eloi Montanez (1)

338. Haploinsufficiency of C9ORF72 selectively impairs autophagy in C9ORF72-linked ALS.

Rim Diab1,2,3, Federica Pilotto1,2,3, Niran Maharjan1,2, Shenyi Jiang4,5, Sabine Liebscher4,5,6, Andrea Salzinger4,5,6, Bhuvaneish T. Selvaraj7,8, Siddharthan Chandran7,8, Smita Saxena1,2*

339. Elucidating the timing of TDP-43 related phenotypes using iPSC-derived motor neurons from TARDBP ALS patients

Melissa Nijs*(1)(2), Adria Sicart Casellas (1)(2), Jimmy Beckers (1)(2), Raheem Fazal (1)(2), Ludo Van Den Bosch (1)(2), Philip Van Damme (1)(2)(3)

340. Dynamic Translational Profile of Stressed iPSC-MNs from C9orf72-ALS Patients by Translating Ribosome Affinity Purification (TRAP)

Yinyan Xu* (1,2), Chaitra Sathyaprakash (1), Krisya Louie (1), Ruxandra Dafinca (1), Jakub Scaber (1), Kevin Talbot (1)

341. MAM lipidome changes associated with TDP-43 dysfunction

Anna Fernàndez-Bernal*(1), Meritxell Martin Gari(1), Natalia Mota-Martorell (1), Pol Andrés-Benito (2), Mònica Povedano (3), Elia Obis (1), Isidro Ferrer (3), Reinald Pamplona (1), Manuel Portero-Otin (1)

342. Gap junctions are functionally enhanced in iAstrocytes derived from C9ORF72 repeat expansion patients

Iris S Pasniceanu*, Jannigje Kok, Allan C Shaw, Cleide Dos Santos Souza, Pamela J Shaw, Laura Ferraioulo, Matthew R Livesey

343. Targeting De Novo Fatty Acid Synthesis as a Therapeutic Strategy to Alleviate Non-cell Autonomous Mechanisms in ALS

Maddi Garciandia-Arcelus (1)*, Andrés Jiménez (1,2), Laura Rodríguez (1), Gorka Gereñu (1,2,3), Jon Ondaro-Ezkurra (1,2), José Ignacio Ruíz-Sanz (3), Roberto Fernández-Torrón (1,4), Juan José Poza-Aldea (4), Javier Riancho (2,5), Raul Domíngez (6), Juan Bautista-Espinal (4), Jesus María Aizpurua (7), Gonzalo González-Chinchón (8,9), Miren Zulaica (1,2), María Begoña Ruiz-Larrea (3), Mónica Povedano (6), Adolfo López de Munain (1,2,4,9,10), Gorka Fernández-Eulate (1,4,11,12), Francisco Javier Gil-Bea (1,2,13,14).

344. Epigenetic analysis on organoids for the study of Amyotrophic Lateral Sclerosis

Eveljn Scarian (1), Matteo Bordoni (1), Camilla Viola (2), Francesca Dragoni (1,2), Rosalinda Di Gerlando (1,2), Luca Diamanti (1), Stella Gagliardi (1), Orietta Pansarasa (1)

345. Cognate microglia – T cell interaction induce neurotoxic T cell function in a fast-progressing C9orf72 ALS animal model

Chao Yang (1), Elisabeth Scharf (1), Steffanie Heindl (2), Leonard Lala (1), Stefan Roth (2, 3), Meike Michaelsen (1), Thomas Arzberger (1, 3, 4, 5), Arthur Liesz (2, 3), Dieter Edbauer (1, 3, 6), Qihui Zhou (1,3) (*)

346. An interaction between synapsin and C9orf72 regulates excitatory synapses and is impaired in ALS/FTD

Claudia S. Bauer (1) (*), Rebecca N. Cohen (1), Francesca Sironi (2), Matthew R. Livesey (1), Thomas H. Gillingwater (3, 4), J. Robin Highley (1), Daniel J. Fillingham (1), Ian Coldicott (1), Emma F. Smith (1), Yolanda B. Gibson (1), Christopher P. Webster (1), Andrew J. Grierson (1), Caterina Bendotti (2), and Kurt J. De Vos (1).

347. Cortical network dysfunction in ALS using task-free magnetoencephalography

Michael Trubshaw(1,2)*, Chetan Gohil(1), Katie Yoganathan(1,2), Evan Edmond(1,2), Malcolm Proudfoot(1,2), Mark Woolrich(1), Martin R. Turner(2)

348. isomiRs - a novel family of molecular biomarkers for ALS prognostication

Yahel Cohen* (1,2),Iddo Maggen (1,2), Nancy - Sarah Yacovzada(1,2), Andrea Malaspina (3), Pietro Fratta (3), Joen Wuu (4), Michael Benatar (4), Eran Hornstein (1,2).

349. Performance of serum neurofilament light chain in a wide spectrum of clinical courses of ALS – a cross-sectional multicenter study

Thomas Meyer (1,2), Erma Salkic (1), Torsten Grehl (3), Ute Weyen (4), Dagmar Kettemann (1), Patrick Weydt (5,6), René Günther (7,8), Paul Lingor (9), Jan Christoph Koch (10), Susanne Petri (11), Andreas Hermann (12,13), Johannes Prudlo (13,14), Julian Großkreutz (15), Petra Baum (16), Matthias Boentert (17), Moritz Metelmann (16), Jenny Norden (1), Isabell Cordts (9), Jochen H. Weishaupt (18), Johannes Dorst (19), Albert Ludolph (19,20), Yasemin Koc (1), Bertram Walter (1), Christoph Münch (1,2), Susanne Spittel (1,2), Marie Dreger (1)*, André Maier (1)*, Péter Körtvélyessy (1,21)*, *contributed equally

350. Ultrasound-mediated blood–spinal cord barrier opening prolongs survival in an ALS mouse model

Anne-Sophie Montero (1,2,3), Ilyes Aliouat* (1,2,3,4), Michael Canney (5), Lauriane Goldwirt (6), Samia Mourah (6), Félix Berriat (4), Christian S Lobsiger (4), Pierre-François Pradat (7), François Salachas (4,7), Gaëlle Bruneteau (7), Séverine Boillée (4), Alexandre Carpentier (1,2,3)

351. Motor system connectivity in ALS: A corticomuscular magnetoencephalography study

Katie Yoganathan (1*), Michael Trubshaw (1), Irene Echeverria-Altuna (2,3), Oliver Kohl (2), Thanuja Dharmadasa (1), Nahid Zokaei (2), Andreas Themistocleous (1), Charlotte Stagg (2), Mark Woolrich (2), Anna C Nobre (2,3), Kevin Talbot (1), Alexander G. Thompson (1) and Martin R. Turner (1).

352. Dysfunction Of Cortical Inhibitory Interneurons In Amyotrophic Lateral Sclerosis

Cristina Benetton* (1), Pierre-François Pradat (1, 2), Véronique Marchand-Pauvert (1), Alexandra Lackmy-Vallee (1)

353. NERVE EXCITABILITY DISENTANGLED: HYPEREXCITABILITY IN ALS IS DRIVEN BY ALTERED SLOW POTASSIUM CHANNEL KINETICS

Diederik Stikvoort Garcia(1), Boudewijn Sleutjes(1), Stephan Goedee(1), Leonard van den Berg(1)

354. Social Cognition Impairment in Amyotrophic Lateral Sclerosis

Sara Kadenšek* (1), Vita Štukovnik (2), Janez Zidar (1), Blaž Koritnik (1)

355. Motor band sign is a specific marker of ALS and corresponds topographically to motor symptoms

Charlotte Zejlon(1,2)*, Stefan Sennfält(2,3), Johannes Finnsson(1,2), Bryan Connolly(1,2), Sven Petersson(1,4), Tobias Granberg(1,2), Caroline Ingre(2,3)

356. Investigating cognitive endophenotypes and presymptomatic cognition in unaffected relatives of familial ALS patients: a longitudinal study

Colm G Peelo* (1,2), Sarah Darcy (1,3), Emmet Costello (1,2), Marie Ryan (1,3), Mark Heverin (1), Orla Hardiman (1,3), & Niall Pender (1,2)

357. Cortical network dysfunction in ALS using task-free magnetoencephalography

Michael Trubshaw(1,2)*, Chetan Gohil(1), Katie Yoganathan(1,2), Evan Edmond(1,2), Malcolm Proudfoot(1,2), Mark Woolrich(1), Martin R. Turner(2)

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