



FILSLAN
Filière de Santé Maladies Rares
Sclérose Latérale Amyotrophique
et Maladies du Neurone Moteur



Association pour la Recherche sur
la Sclérose Latérale Amyotrophique
et autres Maladies du Motoneurone



9TH ALS AND MND RESEARCH MEETING

October 11 and 12, 2023

ICM, Paris

PROGRAM

With the support of



Biogen



SCIENTIFIC AND ORGANISATIONAL COMMITTEE

P.COURATIER (FILSLAN), V.GOUTINES (ARSLA)

H.BLASCO (INSERM, CHU Tours), S.BOILLEE (ICM, Paris), P.CORCIA (INSERM, CHU Tours), P.COURATIER (INSERM, CHU Limoges), C.DESNUELLE (ARSLA), L.DUPUIS (INSERM, Strasbourg), G.LE MASSON (INSERM, CHU Bordeaux), V.PAQUIS-FLUCKLINGER (CNRS, CHU Nice), P.F.PRADAT (INSERM, APHP), C.RAOUL (INSERM, CHU Montpellier).

With the participation of the ARSLA Scientific Council, presidents C.RAOUL and P.F.PRADAT

Presentation code: C = conférence, OC = oral communication, RT = round table, P = poster

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Wednesday 11 October 2023

9:30 - 9:45	Opening: Valérie GOUTINES (ARSLA), Philippe COURATIER (FILSLAN)
9:45 - 10:00	FILSLAN "Training through research" project Award <u>Flavien PICARD</u>
10:00-12:00	SESSION 1: PATHOPHYSIOLOGY OF MOTOR NEURONE DISEASES - Conference: Glial cells at the neuromuscular junction: a new therapeutic target in ALS <u>Richard ROBITAILLE</u> University of Montreal 7 oral presentations (6 x 15 minutes and 1 x 5 minutes)
12:00-12:30	Lunch buffet
12:30-13:30	First poster session
13:30-15:30	SESSION 2: BIOMARKERS AND THERAPIES IN MOTOR NEURONE DISEASES - Conference: New treatments for ALS in 2023 <u>Gaëlle BRUNETEAU</u> ACT4ALS, Paris 7 oral presentations (6 x 15 minutes and 1 x 5 minutes)
15:30-16:00	Break / Poster visit (30 minutes)
16:00-17:30	ARSLA SESSION 6 oral presentations (6 x 15 minutes)
17:30	Conclusion Day 1
From 19:00	FILSLAN cocktail ARSLA Awards Evening Location : Institut Imagine, 24, boulevard du Montparnasse, 75015 Paris

Thursday 12 October 2023

9:00-11:00	SESSION 3: MOLECULAR MECHANISMS OF MOTOR NEURONE DISEASES - Conference: Cryptic splicing: from foe to friend in tackling amyotrophic lateral sclerosis <u>Pietro FRATTA</u> University College London, Londres 6 oral presentations (5 x 15 minutes and 1 x 5 minutes)
11:00-11:15	Break / Poster visit (15 minutes)
11:15-12:15	Conference: Mitophagy failure in Alzheimer's disease: a hope for a diagnostic application and therapeutic targeting <u>Mounia CHAMI</u> Sophia Antipolis, IPMC
12:15-12:45	Lunch buffet
12:45-13:45	Second poster session
13:45-14:00	Announcement of ARSLA awards / Research support actions
14:00-16:00	ROUND TABLE: ARTIFICIAL INTELLIGENCE CONTRIBUTION IN ALS RESEARCH Modeling and predicting the progression of neurodegenerative diseases: application to clinical trial design <u>Stanley DURRLEMAN</u> INRIA, ICM, PRAIRIE ; Paris Harnessing Big Data, Omics, and AI <u>Ahmad AL KHLEIFAT</u> King's College London Machine-learning based on neuroimaging patterns: opportunities and challenges <u>Peter BEDE</u> Trinity College, Dublin
16:00-16:30	Closing / General conclusion: Philippe COURATIER (FILSLAN)

DETAILED PROGRAMME

WEDNESDAY 11 OCTOBER 2023

9:30 | OPENING

- Valerie GOUTINES (ARSLA president)
- Philippe COURATIER (FILSLAN animator)

9:45 | FILSLAN « TRAINING THROUGH RESEARCH » PROJECT AWARD 2022

Unconventional secretion of TDP-43 free aggregates by the ubiquitin-specific protease 19 (USP19)

Flavien PICARD

INSERM U1315, CNRS UMR5261, Institut NeuroMyoGène – Physiopathologie et Génétique du Neurone et du muscle, INMG-PGNM, Lyon, France

10:00 | SESSION 1: PATHOPHYSIOLOGY OF MOTOR NEURONE DISEASES

Moderation: Gwendal LE MASSON and Frédérique RENE

- **C1 Conference : Glial cells at the neuromuscular junction : a new therapeutic target in ALS**
Richard ROBITAILLE
University of Montreal
- **Presentations selected from abstracts submitted on the theme of session 1**
7 oral presentations: 6 x 15 minutes (10 minutes + 5 minutes discussion) and 1 x 5 minutes

OC 1.1 – Deciphering the role of TBK1 in mouse motor neurons and microglial cells, and its implications for ALS/FTD pathogenesis

Lenoël I

Institut du Cerveau - Paris Brain Institute - ICM, Inserm U 1127, CNRS UMR 7225, Sorbonne University, Paris, France, team of Dr. Severine Boillee
Financement ARSLA

OC 1.2 - Specific role of neuronal versus microglial P2X4 receptor in ALS pathogenesis and its potential use as a biomarker

Carracedo S

Univ. Bordeaux, CNRS, IMN, UMR 5293, F-33000 Bordeaux, France
Financement ARSLA

OC 1.3 - Lateral hypothalamus-dependent impairment in ALS

Guillot SJ

University of Strasbourg, INSERM, UMR-S 1118, CRBS, Strasbourg, France

OC 1.4 - Engineered TDP43 condensates with controlled assembly/disassembly as model for studying ALS pathological aggregates in human cells

Combe P

CNRS UMR8640, Department of Chemistry, École Normale Supérieure, PSL University, Sorbonne Université, Paris (France)



OC 1.5 - Cortical sensorimotor integration in patients with ALS

Marchand-Pauvert V

Laboratoire d'Imagerie Biomédicale (LIB), Sorbonne Université, U1146 Inserm, UMR 7371 CNRS, Paris, France

OC 1.6 - Better diagnosing and understanding C9orf72 repeat instability in blood and brain organoids of patients with C9orf72-linked ALS

G. Haase

MPATHY-INS, UMS1106, INSERM & Aix-Marseille University, Marseille, France

Financement ARSLA

OC 1.7 - One-carbon metabolism contribution to development and degeneration in ALS

Hernán-Godoy M

INSERM UMR-S 1118, Central and Peripheral Mechanisms of Neurodegeneration, Centre de Recherche en Biomédecine de Strasbourg, Université de Strasbourg, Strasbourg, France

12:00 | LUNCH BUFFET

12:30 | FIRST POSTER SESSION

Presenters must be present in front of their poster

13:30 | SESSION 2: BIOMARKERS AND THERAPIES IN MOTOR NEURONE DISEASES

Moderation: Emilien BERNARD and Marie-Hélène SORIANI

- **C2 Conference: New treatments for ALS in 2023**

Gaëlle BRUNETEAU

ACT4ALS, Paris

- **Presentations selected from abstracts submitted on the theme of session 2**

7 oral presentations: 6 x 15 minutes (10 minutes + 5 minutes discussion) and 1 x 5 minutes

OC 2.1 - Characterization of a therapeutic approach to deliver scFv targeting TDP-43 pathology in ALS

Al Ojaimi Y

UMR 1253, iBrain, Université de Tours, INSERM, Tours, France

Financement ARSLA

OC 2.2 - Validation of exportin-1 (XPO-1) and mitogen-activated protein kinase kinase 2 (MAP2K2) as molecular drug targets in amyotrophic lateral sclerosis

Parvaz M

Technical University of Munich, School of Medicine, Klinikum rechts der Isar, Department of Neurology; Ismaninger Str. 22, 81675 München, Germany

OC 2.3 - Therapeutic administration of the borna virus x protein by a viral vector AAV10 in a mouse model of ALS

Tournezy J

Neurocentre Magendie INSERM U1215, Université de Bordeaux, France

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OC 2.4 - A better 1-year survival prognosis estimation model for amyotrophic lateral sclerosis using UMAP and regression ridge

Anani T

LIP6, Sorbonne University, Paris, France

OC 2.5 - Quantitative brainstem in amyotrophic lateral sclerosis: implications for predicting non-invasive ventilation needs

Khamaysa M

Sorbonne Université, CNRS, INSERM, Laboratoire d'Imagerie Biomédicale, Paris, France
Financement ARSLA

OC 2.6 - Multimodal automated prediction from quantitative MRI for patient classification and stratification

Matthieu Gilson

Institut de Neurosciences de la Timone, Aix-Marseille University, Marseille

OC 2.7 - Implication of central nervous system barrier impairment in amyotrophic lateral sclerosis: gender-related difference in patients

Alarcan H

Laboratoire de Biochimie et Biologie Moléculaire, CHRU Bretonneau, 2 Boulevard Tonnellé, 37000 Tours, France
UMR 1253 iBrain, Université de Tours, Inserm, 10 Boulevard Tonnellé, 37000 Tours, France

15:30 | BREAK / POSTER VISIT

16:00 | ARSLA SESSION

Organisation/Moderation: Cedric RAOUL and Pierre-François PRADAT (Chairmen of the ARSLA Scientific Council)

- **Selected presentations on ARSLA-funded research projects**
(10 minutes + 5 minutes discussion)

OC A1 – Relationship between diaphragm weakness, taste and smell and food intake in ALS

M. Georges^{1,2}, C. Rabec¹, K. Beauvais³, H. Devilliers⁴, A. Jacquin Picques^{*2,3}, E. Bernard⁵.

Department of Pulmonary Medicine and Intensive Care Unit, Constitutive Reference Center for Rare Pulmonary Diseases, OrphaLung, Dijon-Bourgogne University Hospital, Dijon, France.

Centre des Sciences du Goût et de l'Alimentation, UMR 6265, Centre National de la Recherche Scientifique, Institut National de la Recherche Agronomique, University of Bourgogne Franche-Comté, Dijon, France

OC A2 – Corneal Trigeminothopathy and Amyotrophic Lateral Sclerosis: Myth or Reality?

Raoul Kanav KHANNA

Department of ophthalmology, University Hospital of Tours, France

INSERM UMR 1253 iBrain, Team 2 "Neurogenomics and Neuronal pathophysiology", Tours, France

OC A3 – Transactive response DNA-binding protein 43 is enriched at the centrosome in human cells

Philippe Codron

Univ Angers, Equipe MitoLab, Unité MitoVasc, Inserm U1083, CNRS 6015, SFR ICAT, Angers, France

Neurobiology and neuropathology, University-hospital of Angers, Angers, France

Department of Psychiatry and Neuroscience, University of Laval, Québec City, Qc, Canada

OC A4 – Evaluation of the potential of isoprostanooids to predict ALS progression

Vigor Claire

IBMM, Pôle Chimie Balard Recherche, Montpellier University, CNRS, ENSCM, Montpellier, France / LiNCog-Lille Neuroscience and Cognition, Lille University, INSERM, CHU Lille, Lille, France

OC A5 – Développement d'un panel de biomarqueurs pour la SLA : suivi de la pathologie et de l'efficacité de traitement dans un modèle murin SOD1

Stephanie Duquez, Frederique Rene

Personalised Medicine Centre, School of medicine, Ulster University, UK

INSERM U1118- Université de Strasbourg, France



OC A6 – Detection of Peripherin and misfolded SOD1 as novel biomarkers in ALS

Sylvain Lehmann

INM, Univ Montpellier, INSERM, Montpellier, France

17:30 | CONCLUSION DAY 1

Philippe COURATIER (FILSLAN animator)

FROM 19:30 | FILSLAN COCKTAIL & ARSLA AWARDS EVENING

Location: Institut Imagine

9:00 | SESSION 3: MOLECULAR MECHANISMS OF MOTOR NEURONE DISEASES

Moderation: Luc DUPUIS and Pascal LEBLANC

- **C3 Conference: Cryptic splicing: from foe to friend in tackling amyotrophic lateral sclerosis**
Pietro FRATTA
University College London
- **Presentations selected from abstracts submitted on the theme of session 3**
6 oral presentations: 5 x 15 minutes (10 minutes + 5 minutes discussion) and 1 x 5 minutes

OC 3.1 - The maxomod project: multiomic ALS signatures highlight sex differences, molecular subclusters and the MAPK pathway as therapeutic target

Tzeplaeff L

Rechts der Isar Hospital, Technical University of Munich, Munich (Germany)

OC 3.2 - Identifying motor neuron specific alterations in fus deletion mutant zebrafish model of ALS

Khuljana Mingaj

Imagine Institute, Institut National de la Santé et de la Recherche Médicale (INSERM) Unité 1163,

Translational research for neurological disorders, Paris, France

OC 3.3 - Disrupted spontaneous neural activity in the embryonic SOD1^{G93A} mouse model of amyotrophic lateral sclerosis

Kannantha U

Univ. Bordeaux, CNRS, INCIA, UMR 5287, F-33000 Bordeaux, France

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OC 3.4 – Investigating variants in NUP50 as risk factors for amyotrophic lateral sclerosis

Roman O

INSERM U1118, Université de Strasbourg, Centre de Recherche en Biomédecine de Strasbourg, 1 rue Eugène Boeckel, 67000

Strasbourg

Financement ARSLA

OC 3.5 - TBK1 mutant zebrafish show increased programmed cell death and dysregulation of critical pathways involved in amyotrophic lateral sclerosis (ALS)

Raas Q

Laboratory of Translational Research for Neurological Disorders, Imagine Institute, Université de Paris, INSERM UMR 1163,

75015, Paris, France

OC 3.6 - Heterozygous SPTLC1 p.Leu39del is a major cause of slow-progressing juvenile ALS

Guissart C

Laboratoire de Biochimie et Biologie Moléculaire, CHU Nîmes, Univ. Montpellier, Nîmes, France

11:00 | BREAK / POSTER VISIT

11:15 | C4 CONFERENCE

Moderation: Veronique PAQUIS

Mitophagy failure in alzheimer's disease: a hope for a diagnostic application and therapeutic targeting

Mounia Chami

Sophia Antipolis, IPMC

12:15 | LUNCH BUFFET

12:45 | SECOND POSTER SESSION

Presenters must be present in front of their poster

13:45 | ANNOUNCEMENT OF ARSLA AWARDS /RESEARCH SUPPORT ACTIONS

Valerie GOUTINES (ARSLA Director), Cédric RAOUL and Pierre-François PRADAT (Chairmen of the ARSLA Scientific Council)

ARSLA Scientific Committee Jury :

- Pascal BRANCHEREAU (Bordeaux)
- Caroline ROUAUX (Strasbourg)
- Aude-Marie GRAPPERON (Marseille)
- Emilien Bernard (Lyon)

14:00 | ROUND TABLE: ARTIFICIAL INTELLIGENCE CONTRIBUTION IN ALS RESEARCH

Moderation: Pierre-François PRADAT and Helene BLASCO

Theme situation:

(3 X 20 minutes, questions integrated into the debate + 1 hour discussion)

- **TR1 – Modeling and predicting the progression of neurodegenerative diseases: application to clinical trial design**

Stanley DURRLEMAN

INRIA, ICM, PRAIRIE ; Paris

- **TR2 – Harnessing Big Data, Omics, and AI**

Ahmad AL KHLEIFAT

King's College London

- **TR3 – Machine-learning based on neuroimaging patterns: opportunities and challenges**

Peter BEDE

Trinity College, Dublin

16:00 | CLOSING / GENERAL CONCLUSION

Philippe COURATIER (FILSLAN animator)



POSTER SESSION

P1: Beneficial effect of the environmental enrichment on a transgenic mouse model of FTD-ALS (FUS Δ NLS+/-)

Burgard A

Laboratory for Cognitive and Adaptive Neuroscience – UMR7364/CNRS - University of Strasbourg, FR

P2: Manipulating fus in inhibitory neurons shed light on their contribution to ALS- and FTD-like phenotypes

Lorenc F

INSERM UMR-S 1118, Central and Peripheral Mechanisms of Neurodegeneration, Biomedical Research Centre, University of Strasbourg, Strasbourg (France)

P3: Contribution of neutrophils extracellular DNA Traps to amyotrophic lateral sclerosis pathogenesis

Ritacco C

The institute for Neurosciences of Montpellier, Inserm UMR 1298, University of Montpellier, Saint Eloi Hospital, Montpellier, France.

P4: Dysfunction of motor circuits is key to defects in locomotor behavior in SMA mice

Delestrée N

Center for Motor Neuron Biology and Disease, Depts of Neurology and Pathology & Cell Biology, Columbia University, New York, NY, 10032, USA

P5: Zebra fish: a rapid tool to interpret rare variants in ALS

Arthur Forget

Inserm U 1298, INM, Montpellier, France

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P6: Identification of specific molecular markers of ALS vulnerable motoneurons

Issa Y

The Neuroscience Institute of Montpellier, INM, INSERM UMR1298, University of Montpellier, Montpellier, France

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P7: Propagation and toxicity of super oxide dismutase in amyotrophic lateral sclerosis using human iPSC-Derived motor neuron models

Jost Mousseau C

Institut du Cerveau, ICM, Inserm U1127, CNRS UMR7225, Sorbonne Université, Paris, France.

P8: Role of the sympathetic autonomous system in ALS

Ollivier M

Institut des Neurosciences Cognitives et Intégratives d'Aquitaine, CNRS UMR 5287, Bordeaux, France

Financement ARSLA

P9: Involvement of cerebello-spinal projections in amyotrophic lateral sclerosis

Blanchot C

INSERM UMR1298, Institute of Neurosciences of Montpellier, University of Montpellier, Saint-Eloi Hospital, Montpellier, France

P10: Association Between Brain and Upper Cervical Spinal Cord Atrophy Assessed by MRI and Disease Aggressiveness in Amyotrophic Lateral Sclerosis

El Mendili MM

Aix Marseille Univ, CNRS, CRMBM, Marseille, France

APHM, Hopital de la Timone, CEMEREM, Marseille, France

P11: Homozygous COQ7 mutation, a new cause of potentially treatable distal hereditary motor neuropathy
Jacquier A

CNRS UMR 5261, INSERM U1315, Université Lyon1, INMG, Lyon (France)

Centre de Biotechnologie Cellulaire, CHU de Lyon – Hospices Civils de Lyon (HCL), Bron (France)

P12: Unconventional secretion of misfolded SOD1 and toxicity spreading: a novel therapeutic strategy for amyotrophic lateral sclerosis

Gosset P

The Institute for Neurosciences of Montpellier, Inserm UMR 1051, Univ Montpellier, Saint Eloi Hospital, Montpellier, France

Financement ARSLA

P13: Metabolic reprogramming of regulatory T cells as a therapeutic strategy for amyotrophic lateral sclerosis

Marmolejo-Martínez-Artesero S

The Neuroscience Institute of Montpellier, INM, INSERM UMR1051, Montpellier, France

P14: Preconceived ideas about ALS's medical knowledge: a philosophical approach

Fenoy A

UMR 8011 SND, Initiative Humanités Biomédicales, Sorbonne Université, Paris (France)

P15: Evaluation of the safety and efficacy of the atalante exoskeleton in the rehabilitation of patients with amyotrophic lateral sclerosis

Trad G

Laboratoire d'Imagerie Biomédicale, INSERM U1146, CNRS UMR7371, Sorbonne Université, Paris (France)

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P16: The premodials project: identification of a disease signature for presymptomatic and early ALS

Tzeplaeff L

Department of Neurology, Rechts der Isar Hospital of the Technical University Munich, Munich (Germany)

P17: Widespread Alterations in Fast Amyotrophic Lateral Sclerosis Progressors: A Brain DTI and Sodium MRI Study

El Mendili MM

Aix Marseille Univ, CNRS, CRMBM, Marseille, France

APHM, Hopital de la Timone, CEMEREM, Marseille, France

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P18: Functionnal analysis of genetic variants in amyotrophic lateral sclerosis by studying early markers of neurodegeneration

Bedja--lacona L

UMR 1253, IBrain, Université de Tours, Inserm

Financement ARSLA

P19: A non-pharmacological neuromodulative therapeutic approach for amyotrophic lateral sclerosis; a systematic review

Banos M

Inserm U 1146, CNRS UMR 7371, Laboratoire d'imagerie biomédicale, Sorbonne Université, Paris (France).

P20: Similar changes in glycosphingolipid metabolism in ALS and viral infection are modulated by ambroxol

Spedding M

Spedding Research Solutions, Le Vésinet (France)



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