10e Symposium annuel de la Fondation André-Delambre

Programme / Program

Sclérose latérale amyotrophique : Causes et perspectives thérapeutiques

Amyotrophic Lateral Sclerosis : Causes and Therapeutic Perspectives

Organisateurs / Organizers

Jean-Pierre Julien (Centre de recherche IUSMQ, Université Laval)
Jasna Kriz (Centre de recherche IUSMQ, Université Laval)
Heather Durham (Montreal Neurological Institute)
Deborah Rashcovsky (Montreal Neurological Institute)

19-20 septembre
2014
8:30 - 17:00

Institut et hôpital neurologiques de Montréal
Amphitéatre Jeanne-Timmins
3801, rue University, Montréal, Québec

McGill
Centre universitaire de santé McGill
McGill University Health Centre
neuro
80 years / ans
Montreal Neurological Institute and Hospital
Institut et hôpital neurologiques de Montréal
Institut universitaire en santé mentale de Québec
UNIVERSITÉ LAVAL
FONDATION ANDRÉ DELAMBRE

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2014

8:30 - 17:00

Programme / Program

Chers collègues,
Il me fait plaisir de vous accueillir au Symposium annuel sur la SLA de la Fondation André-Delambre qui célèbre cette année son dixième anniversaire. À cette occasion, nous avons convenu qu'il ait lieu au même endroit que le premier Symposium en 2005, à l'Institut et Hôpital neurologiques de Montréal. En 2004, suite à une levée de fonds fructueuse de sa nouvelle Fondation sur la SLA avec l'aide de Céline Dion et René Angélil, monsieur André Delambre m'avait contacté pour des suggestions sur l'utilisation des fonds pour l'avancement des connaissances sur la SLA. Je lui avais suggéré qu'une des façons d'avoir un impact serait de subventionner un symposium annuel réunissant les meilleurs chercheurs dans le monde. Ainsi, un premier symposium fut organisé en 2005 et compte tenu du succès, la Fondation André-Delambre a continué au fil des ans de parrainer un Symposium annuel sur la SLA. Je tiens donc à remercier cette Fondation ainsi que la Société SLA du Québec pour leur support financier au cours des dix dernières années.

Le symposium doit son succès à son organisation souple, à son style informel et à l'enthousiasme des participants. Cet événement favorise une véritable collégialité entre chercheurs. C'est un lieu de rencontre pour discuter des résultats de la fine pointe de la recherche souvent non publiés et pour stimuler de nouvelles collaborations entre laboratoires de recherche dans le monde. Depuis 10 ans, plus de 240 conférenciers de marque en provenance de 13 pays ont participé au Symposium.
Cette année, les conférenciers invités feront une courte visite à La marche pour la SLA de Montréal. Ce sera une occasion pour les chercheurs d'informer les patients et leurs proches des progrès réalisés en recherche, et c'est aussi une façon d'exprimer notre solidarité dans la lutte contre cette maladie. Pour les patients atteints de la SLA, l'espoir c'est la recherche.
Je vous souhaite un bon symposium.

Jean-Pierre Julien

Dear colleagues,
It is my pleasure to welcome you to the tenth Annual Symposium on ALS of the Foundation André-Delambre. On this occasion, we are meeting at the same location as the first symposium in 2005 at the Montreal Neurological Institute. In 2004, following a successful fundraising of its new Foundation on ALS with the help of Celine Dion and René Angélil, André Delambre had contacted me for suggestions on the use of funds for the advancement of knowledge on ALS. I suggested to him that one way to have an impact would be to subsidize an annual symposium gathering the best researchers in the world. Thus, a first symposium was organized in 2005 and given the success the Foundation André-Delambre continued to sponsor an annual Symposium on ALS over the years. So, we are grateful to this Foundation and the ALS Society of Quebec for their financial support over the past decade.

The symposium's success rests on its flexible organization, its informal style and the enthusiasm of participants. This event promotes a genuine collegiality among researchers. It is a meeting place to discuss the results of cutting-edge research often unpublished and to stimulate new collaborations between research laboratories in the world. For 10 years, more than 240 renowned speakers from 13 countries attended the symposium.
This year's guest speakers will make a short visit to Walk for ALS Montreal. This is an opportunity for researchers to inform patients and their relatives of research progress, and it is also a way to express solidarity in the fight against this disease. For patients with ALS, hope is through research.
I wish you an interesting and productive symposium.

Jean-Pierre Julien

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Friday, 19 September, 2014

08:30 - 08:45 **Welcome address**

Jean-Pierre Julien (Centre de recherche de l'IUSMQ (Université Laval)
Guy Rouleau (Director of Montreal Neurological Institute)

Genetics and mechanisms

Chair: ***Jean-Pierre Julien***

08:45 - 9:15 ***Guy Rouleau***, ALS genetics

09:15 – 9:45 ***Robert Brown*** (University of Massachusetts School of Medicine)
ALS genetics: Further observations on excitotoxicity

09:45 – 10:15 ***Leonard Petrucelli*** (Mayo Clinic, Jacksonville)
Molecular Mechanisms and Therapeutic Approaches for FTD/ALS

10:15 – 10:30 **Coffee break and posters**

10:30 – 11:00 ***Heather Durham*** (Montreal Neurological Institute, McGill University)
Mechanisms underlying dendritic atrophy in ALS

11:00 – 11:30 ***Jeffrey Rothstein*** (Johns Hopkins School of Medicine)
C9orf72: Defect in Nuclear Pore trafficking and development of Corrective Therapeutics

11:30 – 12:00 ***Davide Trotti*** (Jefferson University)
Arginine-rich RAN dipeptides linked to C9ORF72-ALS/FTD form toxic nuclear aggregates that initiate in vitro and in vivo neuronal death

12:00 – 13:30 **Lunch and poster session**

Immunity and glial cells

Chair: ***Jasna Kriz*** (IUSMQ, Université Laval)

13:30 – 14:00 ***Stanley Appel*** (Methodist Neurological Institute, Houston)
T Reg Cells Step Up to the Plate in Lou Gehrig's Disease

14:00 – 14:30 ***Michal Schwartz*** (The Weizmann Institute of Science, Israel)
Orchestration of immunity to support degenerating spinal cord involves effector and regulatory cells, and controlled port of entry

14:30 – 15:00 ***Brian Kaspar*** (Research Institute at Nationwide Children's Hospital, Ohio)
Evaluating the astrocytes, microglia and oligodendrocytes on motor neurons in ALS

15:00 – 15:30 ***Serge Przedborski*** (Columbia University, New York)
Astrocyte toxicity and motor neuron degeneration

15:30 – 15:45 **Coffee break**

Chair: ***Pierre Drapeau*** (CHUM, Université de Montréal)

15:45 – 16:15 ***Jasna Kriz*** (IUSMQ, Université Laval)
Deregulation of innate immune responses in early ALS

16:15 – 16:45 ***Kevin Eggan*** (Harvard University) New ideas about ALS from iPS cells

16:45 - 17:15 ***Jonathan Glass*** (Emory University)
Intraspinal stem cell transplantation for ALS: Phase 2 update

17:15 **Cocktail and poster session**

Vendredi 19 Septembre 2014

08:30 - 08:45 **Mots de bienvenue**

Jean-Pierre Julien (Centre de recherche de l'IUSMQ (Université Laval)
Guy Rouleau (Director of Montreal Neurological Institute)

Génétique et mécanismes

Président: ***Jean-Pierre Julien***
08:45 - 9:15 ***Guy Rouleau,*** ALS genetics
09:15 – 9:45 ***Robert Brown*** (University of Massachusetts School of Medicine)
ALS genetics: Further observations on excitotoxicity
09:45 – 10:15 ***Leonard Petrucelli*** (Mayo Clinic, Jacksonville)
Molecular Mechanisms and Therapeutic Approaches for FTD/ALS

10:15 – 10:30 **Pause café**

110:30 – 11:00 ***Heather Durham*** (Montreal Neurological Institute, McGill University)
Mechanisms underlying dendritic atrophy in ALS
11:00 – 11:30 ***Jeffrey Rothstein*** (Johns Hopkins School of Medicine)
C9orf72: Defect in Nuclear Pore trafficking and development of Corrective Therapeutics
11:30 – 12:00 ***Davide Trotti*** (Jefferson University)
Arginine-rich RAN dipeptides linked to C9ORF72-ALS/FTD form toxic nuclear aggregates that initiate in vitro and in vivo neuronal death

12:00 – 13:30 **Pause midi et visite des affiches**

Immunité et cellules gliales

Président: ***Jasna Kriz*** (IUSMQ, Université Laval)
13:30 – 14:00 ***Stanley Appel*** (Methodist Neurological Institute, Houston)
T Reg Cells Step Up to the Plate in Lou Gehrig's Disease
14:00 – 14:30 ***Michal Schwartz*** (The Weizmann Institute of Science, Israel)
Orchestration of immunity to support degenerating spinal cord involves effector and regulatory cells, and controlled port of entry
14:30 – 15:00 ***Brian Kaspar*** (Research Institute at Nationwide Children's Hospital, Ohio
Evaluating the astrocytes, microglia and oligodendrocytes on motor neurons in ALS
15:00 – 15:30 ***Serge Przedborski*** (Columbia University, New York)
Astrocyte toxicity and motor neuron degeneration

15:30 – 15:45 **Pause café**

Président: ***Pierre Drapeau*** (CHUM, Université de Montréal)
15:45 – 16:15 ***Jasna Kriz*** (IUSMQ, Université Laval)
Deregulation of innate immune responses in early ALS
16:15 – 16:45 ***Kevin Eggan*** (Harvard University) New ideas about ALS from iPS cells
16:45 - 17:15 ***Jonathan Glass*** (Emory University)
Intraspinal stem cell transplantation for ALS: Phase 2 update

17:15 **Cocktail et visite des affiches**

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Saturday, 20 September, 2014

Animal models

Chair: ***Heather Durham*** (Montreal Neurological Institute, McGill University)

08:15 – 08:45 ***J. Alex Parker*** (Université de Montréal)
Role of the innate immune system in motor neuron degeneration in C. elegans ALS models

08:45 – 09:15 ***Ivana Munitic*** (University of Rijeka, Croatia)
A Mouse Model of Optineurin Insufficiency

09:15 – 09:45 ***Janice Robertson*** (Toronto University)
Pathomechanisms of TDP-43 in ALS/FTLD studied in transgenic mice

09:45 – 10:00 **Coffee break**

10:00 – 10:30 ***Pierre Drapeau*** (Université de Montréal)
Restoring neuromuscular transmission in ALS with neuroleptic

10:30 – 11:00 ***Makoto Urushitani*** (Kyoto University)
Molecular epitope identification for misfolded/unfolded TDP-43 in ALS

11:00 – 13:30 **Lunch at Parc Maisonneuve**

Montreal Walk for ALS

Protein misfolding and Experimental therapeutics

Chair: ***Christine Vande Velde*** (CHUM, Université de Montréal)

13:30 – 14:00 ***Neil Cashman*** (University of British Columbia)
SOD1 Propagated Misfolding in ALS: New Findings

14:00 – 14:30 ***Jacob Ian Ayers*** (McKnight Brain Institute, Florida)
Experimental transmissibility of mutant SOD1 motor neuron disease

14:30 – 15:00 ***Steven Perrin*** (ALS Therapy Development Institute, Cambridge)
Development of Therapeutics Targeting Misfolded SOD1

15:00 – 15:30 ***Jean-Pierre Julien*** (IUSMQ, Laval University)
Single chain antibodies for ALS treatment

15:30 – 15:45 **Coffee break**

Chair: ***Angela Genge*** (Montreal Neurological Institute, McGill University)

15:45 – 16:15 ***Timothy Miller*** (Washington University, St-Louis)
Development of Novel Therapies for ALS

16:15 – 16:45 ***Lorne Zinman*** (Toronto University)
Targeted Nuclear Factor Kappa Beta in ALS: A Phase II Clinical Trial.

16:45 – 17:15 ***Angela Genge*** (Montreal Neurological Institute, McGill University)
Hopes and Hurdles for Industry sponsored ALS trials

17:15 **Jean-Pierre Julien / Closing comments**

Samedi, 20 Septembre, 2014

Modèles animaux

Président: ***Heather Durham*** (Montreal Neurological Institute, McGill University)

08:15 – 08:45 ***J. Alex Parker*** (Université de Montréal)
Role of the innate immune system in motor neuron degeneration in C. elegans ALS models

08:45 – 09:15 ***Ivana Munitic*** (University of Rijeka, Croatia)
A Mouse Model of Optineurin Insufficiency

09:15 – 09:45 ***Janice Robertson*** (Toronto University)
Pathomechanisms of TDP-43 in ALS/FTLD studied in transgenic mice

09:45 – 10:00 Pause café

10:00 – 10:30 ***Pierre Drapeau*** (Université de Montréal)
Restoring neuromuscular transmission in ALS with neuroleptic

10:30 – 11:00 ***Makoto Urushitani*** (Kyoto University)
Molecular epitope identification for misfolded/unfolded TDP-43 in ALS

11:00 – 13:30 Lunch au Parc Maisonneuve

Marche de Montréal pour la SLA

Agrégats de protéines et Thérapies expérimentales

Président: ***Christine Vande Velde*** (CHUM, Université de Montréal)

13:30 – 14:00 ***Neil Cashman*** (University of British Columbia)
SOD1 Propagated Misfolding in ALS: New Findings

14:00 – 14:30 ***Jacob Ian Ayers*** (McKnight Brain Institute, Florida)
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14:30 – 15:00 ***Steven Perrin*** (ALS Therapy Development Institute, Cambridge)
Development of Therapeutics Targeting Misfolded SOD1

15:00 – 15:30 ***Jean-Pierre Julien*** (IUSMQ, Laval University)
Single chain antibodies for ALS treatment

15:30 – 15:45 Pause café

Président: ***Angela Genge*** (Montreal Neurological Institute, McGill University)

15:45 – 16:15 ***Timothy Miller*** (Washington University, St-Louis)
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16:15 – 16:45 ***Lorne Zinman*** (Toronto University)
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16:45 – 17:15 ***Angela Genge*** (Montreal Neurological Institute, McGill University)
Hopes and Hurdles for Industry sponsored ALS trials

17:15 Jean-Pierre Julien / Mot de la fin

10e Symposium annuel de la Fondation André-Delambre

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GAIT INITIATION DISORDERS AND BRAKING CAPACITY IN SUBJECTS WITH AMYOTROPHIC LATERAL SCLEROSIS

Mohamed Ouertani M.O
Annabelle Couillandre A.C

Université du Québec à Trois-Rivières
Université de Paris Ouest Nanterre La Défense

Characterize the gait initiation in ALS subjects compared to healthy group, through motor performance (V). Explored postural instability via the braking index.

8 ALS subjects (n = 8, 66 ± 11.7 years), and 8 healthy (n = 8, 65 ± 1.6 1ES years) participated in this study.

The subjects had to perform a self-controlled locomotor task, walking 3 meters and then return to starting point.

The gait was carried out with different speeds: spontaneous,and fast for ALS subjects. Spontaneous, slow and Fast in the group of healthy subjects, with 15 trials for each experimental condition. Only the gait initiation phase (the first step on the platform) was analyzed.

The results obtained shows that at identical speed walking, ALS motor performance is reduced compared to that of healthy subjects. The alteration of the braking process is a further evidence of postural instability, characteristic of some ALS subjects. Exploitation of EMG data to address the concept of motor program.

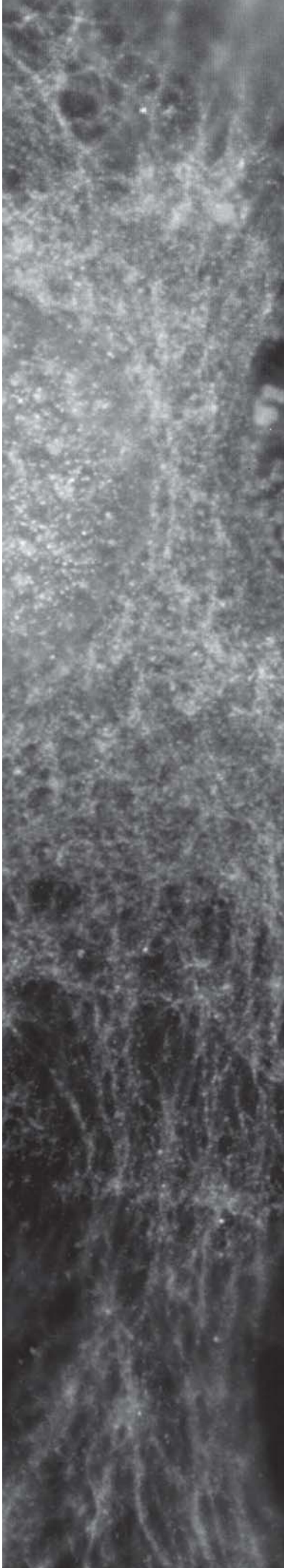
Cerebral structures and biomechanical parameters Relations (fMRI), better understanding of neurophysiology and anatomy at the origin of the GI process.

DÉTECTION PRÉCOCE D'ANOMALIES STRUCTURELLES ET MOLÉCULAIRES DANS UN MODÈLE DE PEAUX RECONSTRUITES PAR GÉNIE TISSULAIRE DÉRIVÉES DE PATIENTS SLA

Bastien Paré M.Sc.[1,2], Lydia Touzel Deschênes M.Sc.[1,2], François-Dominique Scott B.Sc.[1,2], Pierre Provencher[3], Stéphan Saikali MD[4], Peter Gould MD[4], Jean-Pierre Bouchard MD [3], Patrick Dion Ph.D[5], Nicolas Dupré MD, M.Sc.[3], François Berthod Ph.D.[1,2], Guy Rouleau MD, Ph.D.[6] and François Gros-Louis Ph.D.[1, 2]

1. Département de chirurgie, Faculté de médecine, Université Laval, Québec, Canada
2. Centre de recherche du CHU de Québec, axe de médecine regénératrice, LOEX-Hôpital de l'Enfant-Jésus, Québec, Canada
3. Département de sciences neurologiques, CHU de Québec, Hôpital de l'Enfant-Jésus, Québec Canada
4. Unité de neuropathologie, CHU de Québec, Hôpital de l'Enfant-Jésus, Québec Canada
5. Département de pathologie et de biologie cellulaire, Université de Montréal, Montréal, Canada
6. Institut et hôpital neurologique de Montréal, Université McGill, Montréal, Canada

OBJECTIF : La sclérose latérale amyotrophique (SLA) est une maladie neurodégénérative caractérisée par une perte sélective des neurones moteurs. Il est impossible présentement de prédire l,évolution de la maladie puisqu,il n,existe pas de méthode pour la diagnostiquer précocement ni de biomarqueurs de progression. Il a récemment été démontré que plusieurs autres types cellulaires et autres organes situés à l,extérieur du système nerveux, notamment la peau, peuvent aussi être affectés dans la SLA. Dans cette perspective, nous avons été les premiers à développer un modèle in vitro de peau humaine reconstruite en laboratoire dérivée de cellules de patients. Afin de valider ce modèle et dans le but de développer une nouvelle approche diagnostique, nous souhaitons maintenant élargir notre étude afin d,inclure un plus grand nombre de patients. MÉTHODES : Des colorations standards ont été utilisées afin de caractériser les peaux reconstruites dérivées des patients. Des coupes histologiques ont été analysées en microscopie confocale. RÉSULTATS : Ce modèle nous a permis de mettre en évidence plusieurs anomalies structurales et moléculaires présentes uniquement chez les patients. Entre autres, les peaux reconstruites présentent une non-différenciation épidermique, une jonction dermo-épidermique anormale et une désorganisation du collagène. Ces anomalies ont de plus été détectées chez plusieurs patients ne démontant à ce jour aucun symptôme clinique de la maladie. CONCLUSION : La possibilité de modéliser la SLA à partir de cellules somatiques des patients apporte une nouvelle dimension pour la compréhension des mécanismes pathologiques sous-jacents, mais aussi dans le traitement et le diagnostic de cette maladie.



INDUCTION OF NF-KB ACTIVATION BY ALS-LINKED UBIQUILIN-2 MUTANT

Vincent PICHER-MARTEL, Ali AYOUAZ, Daniel PHANEUF and Jean-Pierre JULIEN

CRIUSMQ, Department of Psychiatry and Neuroscience, Quebec, Canada.

Amyotrophic lateral sclerosis (ALS) is the most frequent adult-onset motor disease and is characterized by progressive death of upper and lower motor neurons. This degeneration leads to progressive paralysis of skeletal muscle and, unfortunately, to patient,s death within 2-5 years of symptoms. Most of ALS cases are sporadic (90%) and only 5-10% are familial. In familial cases, some gene has been linked to the pathology like superoxide dismutase 1 (SOD1) (20%), TAR DNA-binding protein (TDP-43), FUS, P62/SQSTM1 or C9ORF72.
Ubiquilin-2 (UBQLN2) plays an important role in ubiquitin proteasome system (UPS) and autophagy by connecting the UPS and ubiquinated protein. Recently, an X-linked mutation in UBQLN2 gene has been discovered in (ALS) familial cases. Approximately twenty mutations have been identified and the main one is P497H. These patients developed cytoplasmic inclusions positive for major proteins implicated in this neurodegenerative disorder and also show UPS impairment. Furthermore, ALS patients without UBQLN2 mutation also express UBQLN2 positive inclusions, supporting an important role of this protein in ALS physiopathology. There is an emerging role of nuclear factor kappaB (NF-κB) in ALS and other neurologic diseases. For example, it has been shown in our lab that TDP-43 upregulation can enhance activation of NFκB.
We used cell cultures to determine if UBQLN2 mutation and accumulation induce NF-κB activation in ALS pathology. Neuro2A cells, neurons derived from mouse, were stably transfected with NF-κB activation luciferase reporter and then with UBQLN2 WT or P497H plasmids for 48 hours. Luciferase activity and western analysis show an increase in NF-κB activation in cells overexpressing UBQLN2 P497H compare to non-transfected and UBQLN2 WT overexpressing cells. These inclusions also seem to be related to NF-κB pathway proteins, which can explain this activation.

REPLICATION STUDY OF MATR3 IN FAMILIAL AND SPORADIC AMYOTROPHIC LATERAL SCLEROSIS

Leblond CS[1,2], Spiegelman D[2,3], Szuto A[2,3], Dionne-Laporte A[2,3], Laurent SA[2,3], Provencher P[4], Dupré N[4], Dion PA[2,3,5] & Rouleau GA[2,3]

1 Department of Human Genetics, McGill University, Montreal (Qc), Canada
2 Montreal Neurological Institute and Hospital, McGill University, Montreal (Qc), Canada
3 Department of Neurology and Neurosurgery, McGill University, Montreal (Qc), Canada
4 The faculty of Medecine, University of Laval, Quebec city (Qc), Canada
5 Department of Pathology and Cellular Biology, University of Montreal, Montreal (Qc), Canada

Amyotrophic lateral sclerosis (ALS) is a devastative neurodegenerative disorder characterized by an extensive loss of motor neurons in primary motor cortex, brainstem and spinal cord. Genetics studies reported a high heritability of ALS and highlighted genetic factors (penetrant genes or risk factors) underlying this condition. Recently, whole-exome sequencing analysis allowed the identification of missense variations altering MATR3 (Matrin 3) in familial ALS. MATR3 was previously associated to distal myopathy 2 and encodes for a nuclear matrix and DNA/RNA binding protein that has been shown to interact with TDP-43 in a RNA-dependent manner. Here, we assessed the MATR3 mutation frequency in French-Canadian ALS cases and showed that MATR3 mutation accounted for 0% and 1.8% in familial and sporadic cases, respectively. Among the mutations identified in sporadic ALS, the splicing mutation c.48+1G>T raised particularly our interest and resulted to the insertion of 24 amino acids in MATR3 protein. To conclude, our findings support the role of MATR3 in ALS and further studies are needed to provide more light about MATR3 proteinopathy.

Acknowledgments : We first would like to thank the patients involved in this study as well as the people of the laboratory of Dr. Rouleau G.A.. We also like to acknowledge the support of the Canadian Institutes for Health Research (CHIR), the ALS division of the Muscular Dystrophy Association (ALS-MDA), the US ALS Association (ALSA) and ALS society of Canada.

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ALS MOTOR NEURON PROTEOME REVEALED BY QUANTITATIVE MASS SPECTROMETRY

Joseph R. Klim, Namrata Udeshi, Luis A. Williams, Jackson L. Sandoe, Tanya Svink, Brandi N. Davis-Dusenbery, Steven A. Carr, & Kevin Eggan

Department of Stem Cell and Regenerative Biology, Harvard University, the Stanley Center for Psychiatric Research,
The Broad Institute of Harvard and MIT, Cambridge, MA 02138, and the Howard Hughes Medical Institute, USA.

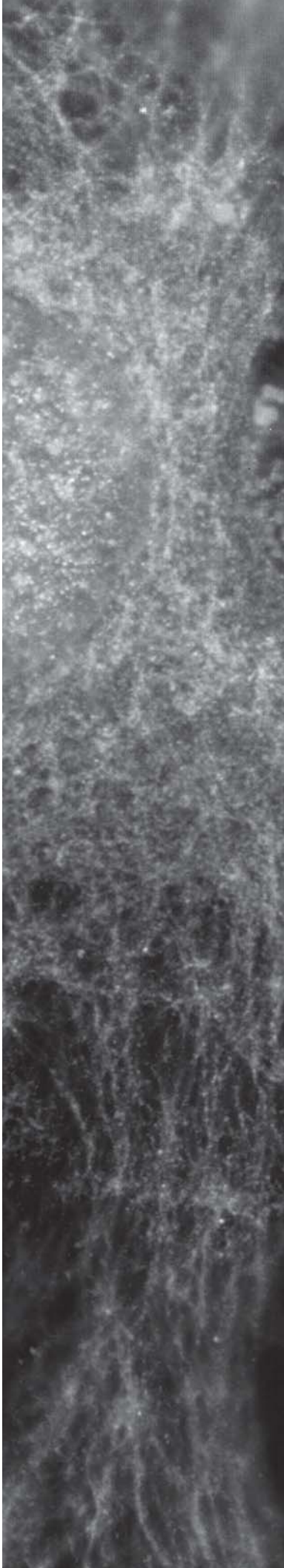
Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disease characterized by the selective loss of upper and lower spinal motor neurons. Our inability to isolate these faulty cells from patients and study them in vitro has impeded progress for decades. To identify the root cause of ALS and develop effective therapeutics, research efforts are needed that drive at the cellular basis of neural degeneration. To this end, our group has introduced an ALS-causing mutation into a human embryonic stem cell line. Specifically, zinc-finger nucleases were used to induce a dominant missense mutation (A4V) in the superoxide dismutase 1 gene (SOD1). Both cell lines can be differentiated into spinal motor neurons with high efficiencies. Using this isogenic pair of stem cell lines, we applied in-depth mass spectrometry-based proteomics to purified populations of stem cell-derived motor neurons. To facilitate relative quantification of proteins from the isogenic samples, stable isotope labeling by amino acids in cell culture (SILAC) was employed. We demonstrate that effective incorporation of the heavy amino acids can be achieved during the differentiation process. Moreover, these experiments resulted in the identification of more than 6000 proteins expressed by motor neurons. Quantitative analysis revealed differences in proteins levels between the neurons with wildtype SOD1 and those with mutant SOD1. These investigations provide clues as to the molecular pathology of ALS and could yield new therapeutic targets.

PRESYMPTOMATIC ALTERATION OF GLIAL CELL FUNCTION AT THE NMJ OF A FAST-TWITCH MUSCLE MAY IMPACT REINNERVATION IN THE SOD1G37R MOUSE MODEL.

Martineau, E.[1,2],Robitaille, R.[1.2]

1. Département de physiologie, Université de Montréal, Montréal, Quebec, H3C 3J7 Canada, and
2. Groupe de recherche sur le système nerveux central, Université de Montréal, Montréal, Quebec H3C 3J7, Canada

Glial cells are known to play a major role in the progression of Amyothrophic lateral sclerosis (ALS). However, the contribution of Perisynaptic Schwann cells (PSCs), glial cells at the neuromuscular junction (NMJ), is still ill-defined in ALS despite that they regulate both the synaptic and structural plasticity of the healthy NMJ. We previously reported changes in PSC properties at a presymptomatic and pre-onset stage in the slow-twitch Soleus muscle. Since fast-twitch muscles are more vulnerable in ALS, we tested whether PSC properties were also altered in the fast-twitch Sternomastoid muscle. Using Ca2+ imaging on isolated nerve muscle preparations, we found that glial Ca2+ responses to neurotransmitter release evoked by motor nerve stimulation were greatly diminished at P180. Furthermore, PSCs on a single NMJ displayed heterogeneous responses suggesting that PSCs are unable to accurately decode synaptic activity. Since adequate PSC decoding is required for proper maintenance of the NMJ, these presymptomatic glial abnormalities suggest that PSCs may not respond adequately to denervation in ALS. Consistent with this possibility, PSCs at denervated NMJs of end-stage animals failed to upregulate Mac-2, a marker of glial activation, while PSCs at innervated NMJs upregulated it. Furthermore, fewer PSC processes at denervated NMJs and nerve terminal sprouting at innervated NMJs were observed. Interestingly, these defects were less pronounced in the Soleus muscle which is consistent with the partial resistance of slow-twitch muscles. Together, these results show that neuron-glia communication is altered early in ALS which may result in a disorganised glial response to denervation.



INVESTIGATING THE ROLE OF THE NBAF COMPLEX IN ALS

Tibshirani M., Durham H.D.

Montreal Neurological Institute, McGill University

Introduction: Dendritic attrition of motor neurons is a common pathological finding in ALS, however while maintaining dendritic connections is important, the mechanisms by which dendritic attrition occurs is understudied. The hnRNPs FUS and TDP43 accumulate in the cytoplasm of motor neurons in ALS patients and are involved in similar RNA-processing pathways. Brg1 is part of a multisubunit complex known as the neuronal Brg1-associated factor (nBAF) chromatin remodeling complex. The nBAF complex plays a role in neuronal differentiation and dendritic outgrowth, which depend on its subunit composition. In this study, we examined the effect of cytoplasmic accumulation of FUS and TDP43 on dysregulation of the nBAF complex relating to changes in dendritic morphology.

Methods: Motor neurons in dissociated spinal cord cultures prepared from E13 mouse embryos were microinjected with plasmids encoding either empty vector, WT or mutant FUS or TDP43 along with plasmid encoding mCherry to visualize cell morphology. After 3 days, dendritic morphology was analyzed with the MATLAB program Bonfire.

Results: Significant dendritic attrition occurred in motor neurons expressing either ALS-mutant FUS or TDP43 compared to their WT counterparts. The nBAF components, Brg1 and BAF53b, were significantly depleted from the nucleus when either FUS or TDP43 accumulated in the cytoplasm and dendritic attrition was prevented by co-overexpression of Brg1. Treatment of motor neurons with the HDAC inhibitor SAHA maintained Brg1 in the nucleus in the presence of mutant FUS/TDP43 and maintained dendritic architecture.

Conclusion: Our study links the cytoplasmic accumulation of FUS and TDP-43, which occurs in familial and sporadic forms of ALS, to epigenetic changes including disruption of nBAF complex activity, leading to retraction of the dendritic arbor.

Acknowledgments : The authors thank Sandra Minotti for the spinal cord-DRG cultures.

DELETERIOUS VARIATIONS IN THE ESSENTIAL MRNA METABOLISM FACTOR, HGLE1, IN ALS PATIENTS

Kaneb H.M.[1,2,3], Folkmann A.W.4, Belzil V.V.[3,5], Jao L-E.[4], Leblond C.S.[1,6], Girard S.L.[3,7], Daoud H.[3,8], Noreau A.[3,9], Rochefort D.[1], Hince P.[1], Szuto A.[3], Vidal S.[10], André-Guimont C.[11], Camu W.[12], Bouchard J-P.[13], Dupré N.[13], Meininger V.[14], Rouleau1 G.A.[2], Wente S.R.[4], Dion P.A.[1,2,3,9]

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ALS is a fatal neurodegenerative disorder characterised by the selective death of motor neurons. Causative mutations in the global RNA processing proteins TDP-43 and FUS as well as their aggregation in ALS patients have identified defects in RNA metabolism as a salient feature in this disease. Lethal congenital contracture syndrome 1 (LCCS1) and lethal arthrogryposis with anterior horn cell disease (LAAHD) are autosomal recessive fetal motor neuron diseases that are caused by mutations in another global RNA-processing protein, hGle1. In this study we carried out the first screening of GLE1 in ALS patients (173 familial and 760 sporadic) and identified two deleterious variations (one splice site and one nonsense) and two missense variations. Functional analysis of the deleterious variations revealed them to be unable to rescue motor neuron pathology in zebrafish morphants lacking Gle1. Furthermore, in HeLa cells both variants caused a depletion of hGle1 at the nuclear pore where it carries out an essential role in nuclear export of mRNA. These results suggest a haploinsufficiency mechanism and point to a causative role for GLE1 mutations in ALS patients. This further supports the involvement of global defects in RNA metabolism in ALS.

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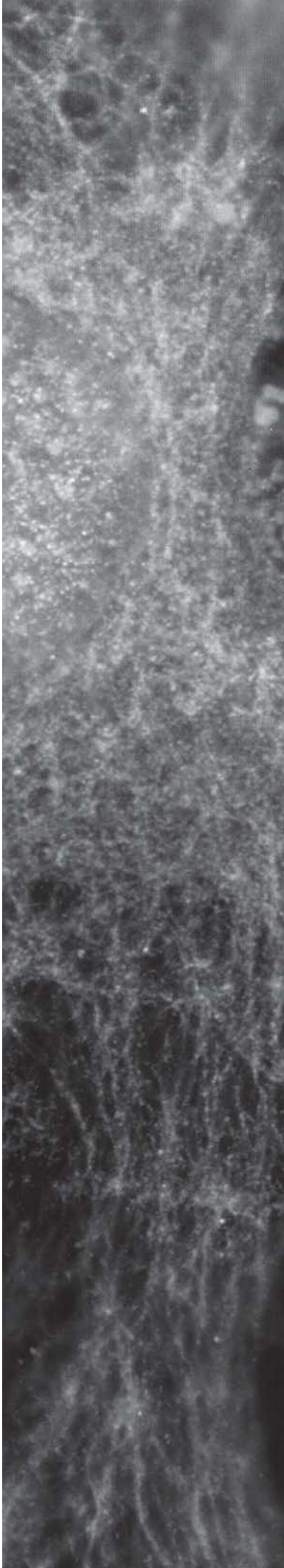

10e Symposium annuel de la Fondation André-Delambre

19-20 septembre 2014

8:30 - 17:00

MOUSE MODEL FOR CELL-TYPE SPECIFIC MICROGLIA MOLECULAR PROFILE ANALYSIS IN ALS.

Béland, L.-C., Boutej, H., Gravel, M., Lalancette-Hébert, M., Kriz, J.

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Previous study has shown that microglosis in late-stage ALS in spinal cord differs greatly from microgliosis in other CNS tissue, from microgliosis before the ALS onset and from microglia activated with LPS. Generating a transgenic mice model for cell-type specific molecular profile analysis of microglia and using translating ribosome affinity purification would allow us to determine what makes the difference in the activation of lumbar spinal cord microglia in late-stage ALS at the proteomic level.

FUS/FUST-1 IS INVOLVED IN OSMOTIC STRESS RESPONSE

Therrien, Martine[1], Rouleau, Guy A.[2], Dion, Patrick A.[2,3], Parker, J. Alex[1,4]

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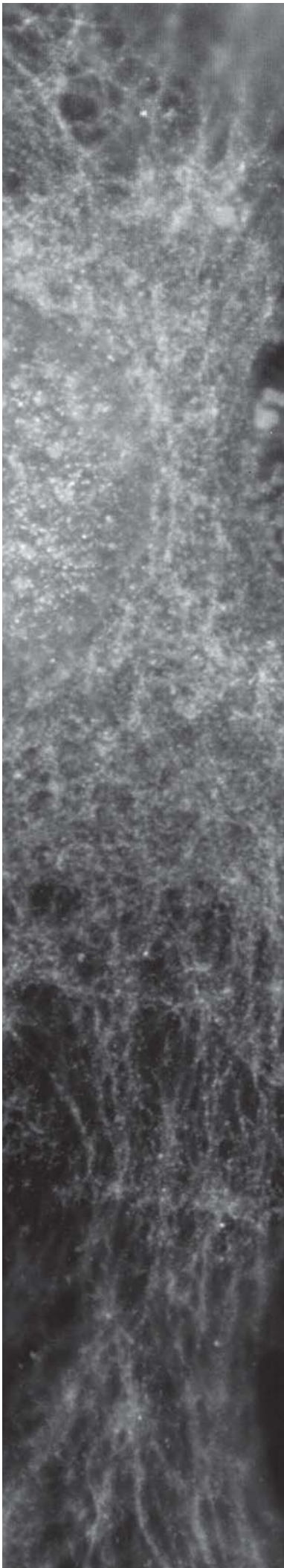
Many proteins involved in amyotrophic lateral sclerosis (ALS) are evolutionarily conserved in the worm Caenorhabditis elegans. C. elegans is a transparent nematode widely used for anatomical, behavioural and genetic studies. It possesses an invariant cell lineage that include in the adult nematode 302 neurons. Also, cellular stress responses and survival mechanisms are genetically regulated and conserved from the nematode and human. Therefore, our group, and others, has used C. elegans to model different aspects of neurodegenerative diseases including ALS.

In this project, we aim to understand the function of FUS and its C. elegans orthologue fust-1. Using a deletion mutant worm, fust-1(tm4339), we have shown that a decreased expression of fust-1 causes motility impairment and fust-1 is required for the proper function of the insulin/IGF pathway to regulate lifespan. Finally, FUST-1 is involved in the normal response to osmotic stress, where a decreased expression of fust-1 causes a hypersensitivity to osmotic stress and osmotic stress induces the expression of FUST-1.

Cellular osmotic stress can have many cellular consequences including molecular crowding and abnormal protein aggregation. The relation between FUS and osmotic stress should be further investigated and could help understand the link between the stress response pathways, neurodegeneration and the different ALS genes.



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THE E3 UBIQUITIN LIGASE TRAF6 STIMULATES MUTANT SOD1 AGGREGATION AND INTERACTS WITH MITOCHONDRIAL SURFACE-ASSOCIATED MISFOLDED SPECIES.

Semmler, S.[1,3], Pickles, S.[3,4], Destroismaisons, L.[3], McBride, H.[1,2], Vande Velde, C.[3,4,5]

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Amyotrophic lateral sclerosis (ALS) is a progressive and ultimately fatal neurodegenerative disease, characterized by the loss of motor neurons in the spinal cord and consequent paralysis of skeletal muscles. Mutations in superoxide dismutase 1 (SOD1) account for 20% of familial and about 3% of sporadic ALS cases and mitochondria have long been proposed as targets for toxicity. Mutant SOD1, adopting an aberrant protein folding pattern, preferentially binds to the outer face of mitochondria in patient-derived cell lines and in an age-dependant manner in rodent models of ALS. The presence of misfolded SOD1 on mitochondria correlates with abnormal mitochondrial morphology, dysfunctional mitochondrial protein import and excessive production of superoxide. However, the exact mechanism of misfolded SOD1-mediated mitochondrial dysfunction and motor neuron toxicity remains unexplained.

In a screen for interacting partners of mitochondrial-associated misfolded SOD1 in the SOD1G93A rat model, we identified the E3 ubiquitin ligase TNF-receptor associated factor 6 (TRAF6). TRAF6 is already reported to play a role in various neurodegenerative diseases, including Alzheimer,s, Parkinson,s and Huntington,s disease, where it frequently ubiquitinates disease-associated mutant proteins and engages in their accumulation into potentially toxic aggregates. We hypothesize that TRAF6 is also involved in the ubiquitination and aggregation of mutant SOD1 on mitochondria and thereby contributes to mitochondrial malfunction and the degeneration of motor neurons in ALS.

Here, we show that misfolded SOD1 interacts in a conformation-dependent manner with TRAF6 on spinal cord mitochondria of symptomatic SOD1G93A rats. TRAF6 is not transcriptionally upregulated, but rather recruited to the mitochondrial surface with disease progression. Interestingly, the immunoprecipitate of the TRAF6-interacting misfolded SOD1 conformer on mitochondria is polyubiquitinated and in an aggregated state. In culture, we demonstrate that the knockdown of TRAF6 alleviates mutant SOD1 aggregate formation and that, reversely, TRAF6 overexpression exacerbates it. To establish relevance for TRAF6 in the cell type primarily lost in ALS, we find TRAF6 predominantly expressed in lumbar spinal motor neurons.

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OPPOSITE SYNAPTIC ALTERATIONS AT THE NEUROMUSCULAR JUNCTION IN AN ALS MOUSE MODEL : WHEN MOTOR UNITS MATTER

Tremblay, E., Martineau, É., Robitaille, R.

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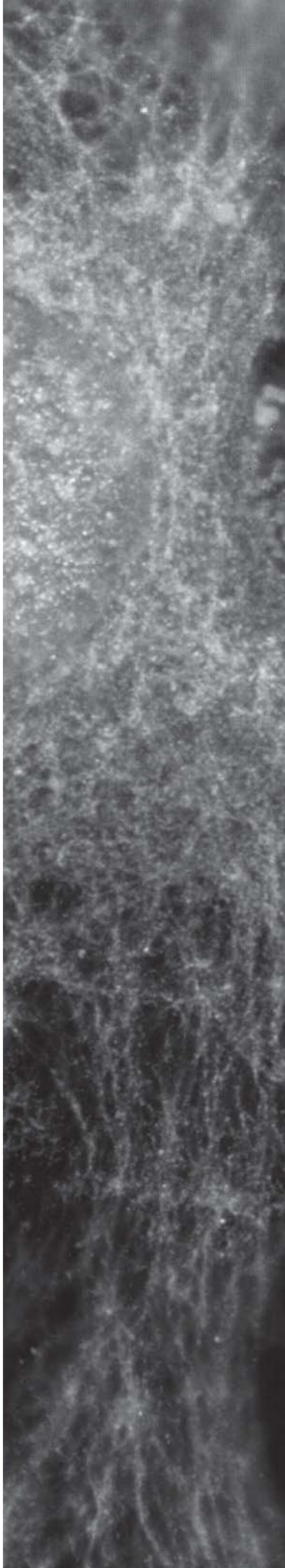
Amyotrophic lateral sclerosis is a late-onset neurodegenerative disease that leads to paralysis and death in 2 to 5 years after the diagnosis. The loss of the neuromuscular junction (NMJ) is the first event in the disease process. NMJs show temporal patterns of denervation in ALS depending on the motor unit (MU) types: NMJs from fast-fatigable (FF) MUs are the first to denervate, followed by the fast resistant (FR) and finally the slow (S) MUs. However, no study has taken into account the MU types when investigating neurotransmission at the NMJ in ALS. We therefore hypothesized that NMJ function would be altered in a MU type specific manner. We used electrophysiology and immunohistochemistry to study synaptic activity in two nerve-muscle preparations of the SOD1G37R mouse and their wild-type (WT) littermates: the slow-twitch Soleus and the fast-twitch extensor digitorum longus (EDL). At a presymptomatic stage (P140), synaptic strength was already altered : the FF synapses of the EDL and the S synapses of the Soleus had respectively a lower and a higher quantal content compared to WT. Long-term synaptic plasticity was also reduced in the EDL. At a symptomatic age (P380), differences in quantal content were still presents in both muscles on innervated NMJs. These oppositely directed changes demonstrate for the first time how NMJs physiology in ALS can be linked to their selective vulnerabilities. This study provides insights for a better understanding of NMJ function during the disease that is essential to the development of a proper treatment at the NMJ in ALS.

Acknowledgments : FRSQ, ALS CANADA, FCI, CIHR



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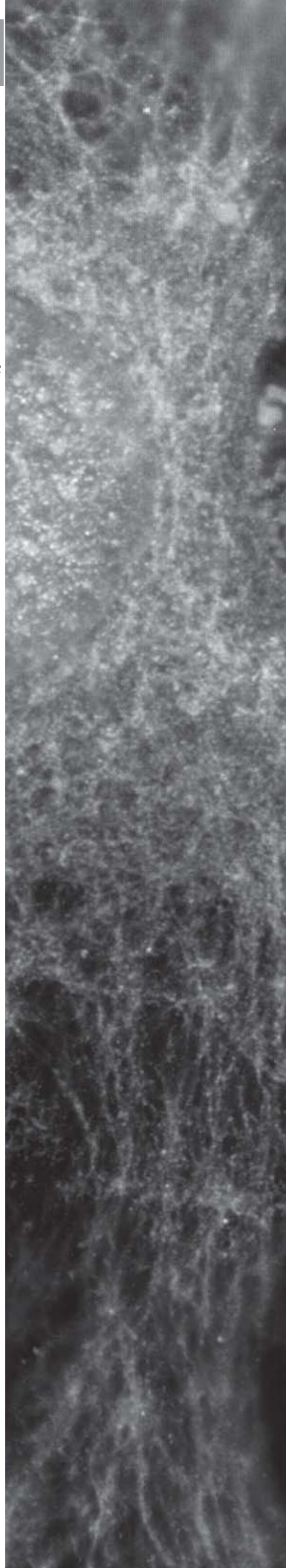

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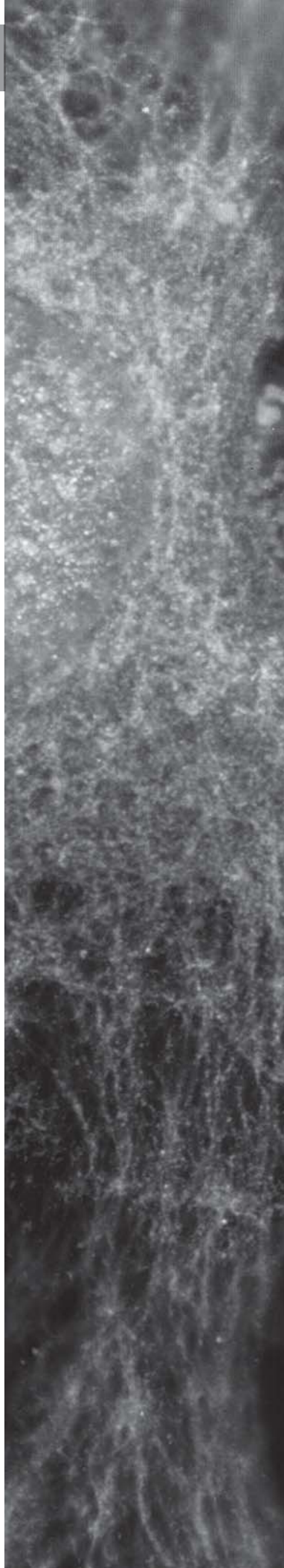

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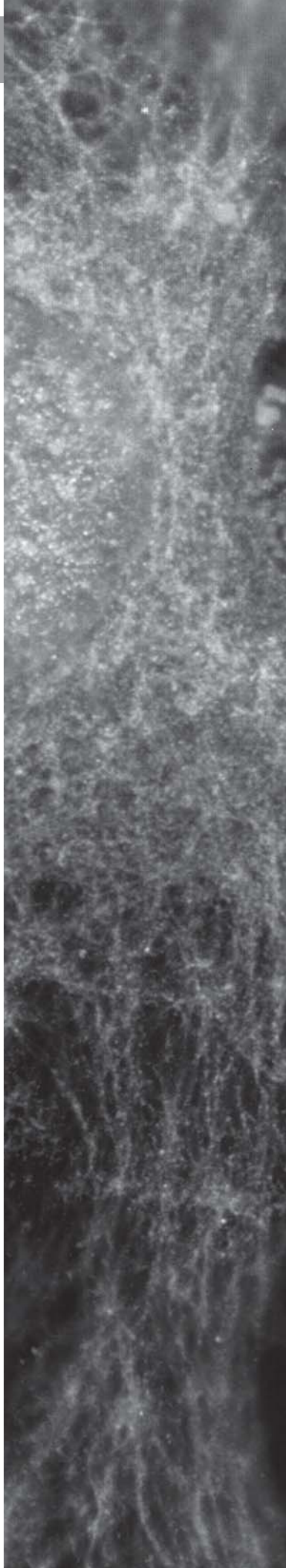

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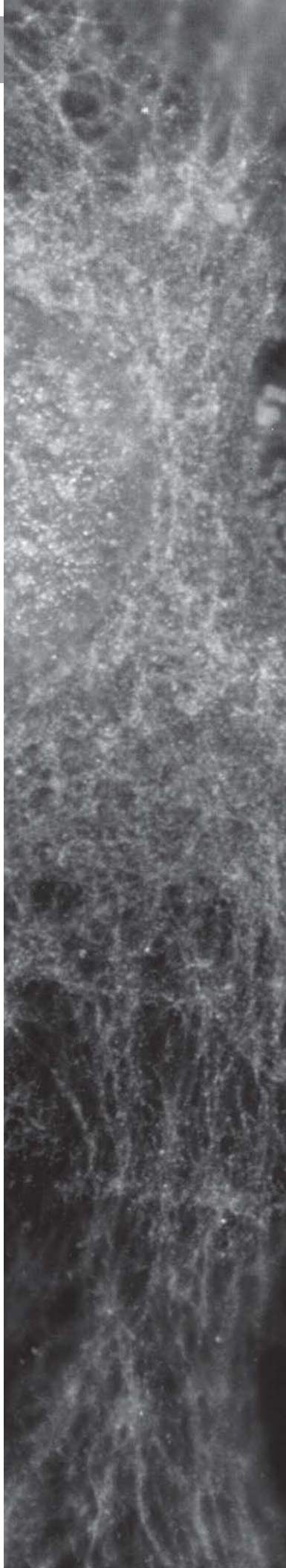

10e Symposium annuel de la Fondation André-Delambre

19-20 septembre 2014

8:30 - 17:00

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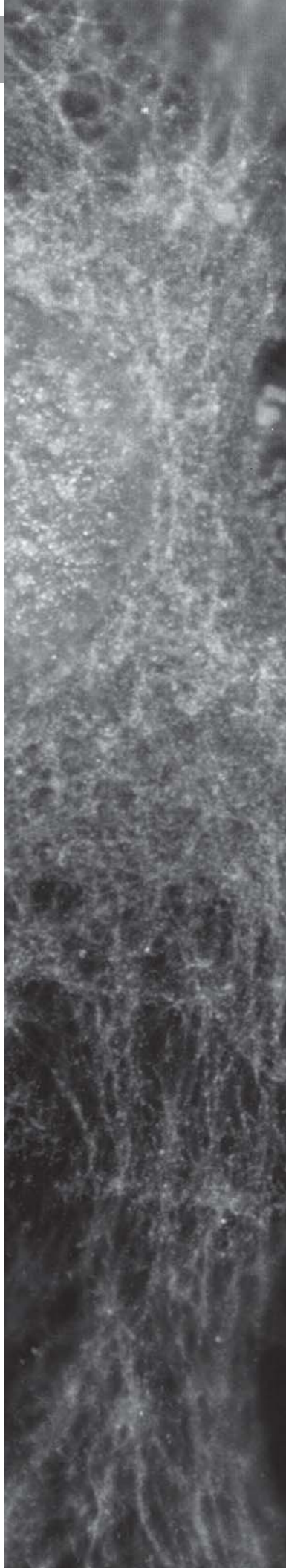

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