SUMMARY OF ALS RESEARCH IN CANADA

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The ALS Society of Canada is committed to:

- Investing our research funds where they will have the most impact.
- Funding excellent and relevant peer-reviewed research.
- Funding research that is evaluated at a high level using international evaluation methods adopted by the Canadian Institutes of Health Research.

We fund only the most promising projects by the finest scientists and, over the years, this strategy has been rewarded with significant advances in our knowledge of ALS. With the acceleration of results due to advances in neurology and other areas of science, we know that, effective therapies and a cure are now, more than ever, within reach. The following is a list of researchers who are currently studying ALS and neuromuscular diseases in Canada.

Research Projects funded by the ALS Society and/or the Neuromuscular Research Partnership

Neuromuscular Research Partnership

In 1999, The ALS Society of Canada partnered with the Canadian Institutes of Health Research, and Muscular Dystrophy Canada to create the Neuromuscular Research Partnership. This partnership was created to collectively fund health research by providing operating grants in the area of neuromuscular diseases with a mandate to find a cause, treatment options and eventually result in a cure.

Dr. Jean-Pierre Julien

Where:	Université Laval
Project:	Role of Chromogranin-mediated Secretion of Superoxide Dismutase Mutants in ALS Pathogenesis. Chromogranins are molecules made by nerve cells that usually aid production and packaging of other proteins.
Funding:	ALS Society of Canada and The ALS Association in the U.S. - commenced in 2005
Project:	Pathogenic mechanisms associated with neurofilament disorganization
Funding:	NRP (2005) - Five years
Project:	The role of inflammation in pathogenesis of ALS (with Dr. Serge Rivest)
Funding:	NRP (2003)-Five years
Project:	Generation and analysis of a new mouse model for ALS
Funding:	NRP (2002)-One year
Project:	The role of inflammatory cytokines in pathogenesis of ALS
Funding:	NRP (2000)-Three years
Overview:	Developing a novel mouse model for the juvenile form of ALS.

Dr. George Karpati & Dr. Joséphine Nalbantoglu

Where:	McGill University, Montreal Neurological Institute
Project:	Molecular therapies for dystrophin deficiency
Funding:	NRP (2005) - Three years
Project:	Extrasynaptic endogenous utrophin upregulation in skeletal muscle: A therapeutic approach for Duchenne muscular dystrophy (DMD)
Funding:	NRP (2004)-Three years (George Karpati)
Project:	Development of efficient and safe gene transfer to skeletal muscle for the therapy of dystrophin deficiency
Funding:	NRP (2002)-Three years (George Karpati & Josephine Nalbantoglu)
Project:	Utrophin upregulation in skeletal muscle: A Therapeutic Approach for DMD
Funding:	NRP (2000) - Three years
Overview:	Exploring how utrophin, a protein, can be "upregulated" or inserted to lessen the impact of a dystrophin deficiency. DMD is caused by a deficiency of the dystrophin protein.

Dr. David J. Picketts

Where:	Ottawa Health Research Institute
Project:	Genetic dissection of ISWI function during neurogenesis
Overview:	Studying two proteins - SNF2H and SNF2L - for their role in the complex chain of molecules and signals that help to turn stem cells into neurons in a developing embryo to better understand their role in the growth and differentiation of both neurons and muscles.
Funding:	NRP (2005) - Three years

Dr. Michael Sinnreich

Where:	McGill University, Montreal Neurological Institute
Project:	Modular flexibility of dysferlin-possible applications for gene therapeutic strategies
Overview:	Studying unique cases of LGMD 2B (limb-girdle muscular dystrophy) to further understand dysferlin (muscle protein thought to be involved in the repair of muscle damage caused naturally through exercise) and uncover ways to replace its relatively large gene inside the cell.
Funding:	NRP (2005) - One year

Dr. Kenneth E. Hastings

Where:	Montreal Neurological Institute, McGill University
Project:	Fiber-type-specific and actively-regulated gene expression in fast skeletal muscle
Overview:	Investigating the molecular events that determine muscle fiber-type. Muscle fiber is an important aspect of muscle function. Muscle fibres develop into their two separate varieties - fast and slow.
Funding:	NRP (2005) - Three years, NRP (2002)-Three years

Dr. Hakima Moukhles

Where:	University of British Columbia
Project:	Dystroglycan function in glial cells
Overview:	Studying why there is sometimes an effect on the brain when someone has a neuromuscular disorder. Studying the protein dystroglycan implicated in several forms of muscular dystrophy. Glial cells form a protective insulating layer around nerve cells.
Funding:	NRP (2005)- Three years

Dr. Louise R. Simard

Where:	Hôpital Sainte-Justine(Montreal)
Project:	Characterization of "survival of motor neurons" (SMN) gene regulation
Overview:	Studying how SMN gene expression is regulated during development. Mutations in the SMN gene are responsible for childhood spinal muscular atrophy, a severe lower motor neuron disease.
Funding:	NRP (2005) - Three years, NRP (2001) - Three years

Dr. Heather Durham

Where:	McGill University, Montreal Neurological Institute
Project:	The role of protein chaperones and proteasome-mediated proteolysis in the pathogenesis of motor neuron diseases
Overview:	The Durham lab studies why motor neurons are more vulnerable to damage in ALS than other cells in order to identify protective therapies. Durham's lab is investigating the importance of protein chaperones and proteosomes in helping cells deal with damaged proteins. The chaperones' job is to round up damaged proteins (in this case, the mutant SOD1) and shuttle them to the proteosomes where they are chopped up and discarded.
Funding:	NRP (2005)-Five years, NRP (2002)-Three years

Dr. Bernard Brais

Where:	Centre Hosp. de l'Université de Montreal
Project:	Cloning and characterising the mutated gene responsible for a new form of French-Canadian recessive spastic ataxia
Funding:	NRP (2005) - Three years
Project:	Cloning and characterising the mutated gene responsible for a French-Canadian recessive ataxia associated with severe polyneuropathy
Funding:	NRP (2003) - Three years
Project:	Oculopharyngeal muscular dystrophy (OPMD) and polyalanine toxicity
Funding:	NRP (2001)-Three years
Overview:	Seeking to understand how the mutation - a lengthening of a portion of specific protein - leads to OPMD and other related disorders so that further research can be directed toward effective therapies. OPMD is a hereditary disease that results in generalized muscle weakness, particularly those responsible for eyelid elevation and swallowing.

Dr. F.J. Dilworth

Where:	Ottawa Health Research Institute
Project:	Elucidating the mechanisms directing temporally ordered gene expression by myoD
Overview:	Studying how and why the the myoD gene (that codes for the myoD protein molecule) assists in changing the natural stem cells found in the embryo into a highly organized system of skeletal muscles.
Funding:	NRP (2005) - Five years

Edor Kabashi, PhD Student

Where:	McGill University, Montreal Neurological Institute (Dr. Heather Durham's Lab)
Project:	Problems with Protein Disposal in ALS
Overview:	Contributing to a better understanding of pathophysiology of ALS and other neurodegenerative disorders and to offer new therapeutical agents to help people with ALS.
Funding:	Awarded two years of funding by the ALS Society of Canada (2004)

Miranda Tradewell, PhD Student

Where:	McGill University, Montreal Neurological Institute (Dr. Heather Durham's lab)
Project:	The Role of Calcium in Motor Neuron Disease
Overview:	Seeking not only to examine the cause of motor neuron diseases such as ALS, but to gain a better understanding of why the biology of motor neurons makes them vulnerable to the toxicity of mutant proteins compared to other cells.
Funding:	Awarded three years of funding by the ALS Society of Canada (2004)

Dr. Avijit Chakrabartty

Where:	University Health Network Ontario Cancer Institute
Project:	Protein misfolding and conformational disease
Overview:	Studying how protein folding affects cells and the entire body to better understand ALS.
Funding:	NRP (2004) - Three Years

Dr. Robin J. Parks

Where:	Ottawa Health Research Institute Ottawa Hospital Research Institute
Project:	Adenovirus vectors for gene therapy of muscle
Overview:	Has several advantages over traditional Ad vectors and has shown promising results in pre-clinical studies
Funding:	NRP (2004)-Three years (with Jonathan L. Bramson) NRP (2001)-Three years

Dr. Vanessa J. Auld

Where:	University of British Columbia
Project:	Glial cell development and function at the Drosophila neuromuscular junction
Overview:	Studying the physiological basics of glia cells in fruit fly development. Nervous system development and function is guided by control cells called glia.
Funding:	NRP (2004)-Three years

Dr. Jérôme Frenette

Where:	Université Laval
Project:	Inflammatory cell recruitment and function in skeletal muscles following hind limb unloading and reloading: New strategies to prevent muscle atrophy and dysfunction
Overview:	Studying how a side effect of mobility - secondary atrophy - can be reduced or diminished.
Funding:	NRP (2004)-Three years

Dr. Jiming Kong

Where:	University of Manitoba
Project:	ALS: Role of BNIP3 in mutant SOD1-induced motor neuron death
Overview:	Examining ways to slow or decrease ALS symptoms. BNIP3 is a recently discovered protein, which appears to slow and control apoptosis (programmed cell death is a genetically controlled mechanism for the body to eliminate cells that have outlived their usefulness) in motor neurons.
Funding:	NRP (2004)-Three years

Dr. Susan O. Meakin

Where:	Robarts Research Institute (London)
Project:	Nesca, a novel intracellular signaling adapter facilitates neurotrophin dependent neurite outgrowth.
Overview:	Determining the mechanism(s) which facilitate a novel intracellular protein that Meakin and her group have identified, named Nesca -- new molecule containing an SH3 domain at the carboxyterminus.
Funding:	NRP (2004)-Two years

Dr. Robert G. Korneluk

Where:	Children's Hospital of Eastern Ontario (Ottawa)
Project:	The X-linked inhibitor of apoptosis (XIAP): A therapeutic agent for the treatments of muscular dystrophy
Overview:	Helping to develop therapies for neuromuscular disorders. Apoptosis - or programmed cell death - is a genetically controlled mechanism for the body to eliminate cells that have outlived their usefulness.
Funding:	NRP (2004)-Three years

Dr. Janice Robertson

Where:	The Centre for Research in Neurodegenerative Diseases at the University of Toronto
Project:	Peripherin abnormalities in ALS
Overview:	Focusing on peripherin, a key protein involved in the pathological aggregates found in motor neurons in ALS. Peripherin is found in the neurofilament aggregates that clog motor neurons in ALS. By searching for toxic mutations in peripherin, she hopes to discover the echanisms that cause ALS. Robertson is the only researcher in Toronto who is focused exclusively on ALS.
Funding:	NRP (2003) - Three years

Janka Hegedus, PhD Student

Where:	University of Edmonton, Alberta. Works in Dr. Tessa Gordon's lab
Project:	Thesis work involves characterizing the progressive loss of motoneurons and their muscle fiber connections in ALS.
Funding:	The NRP, National Sciences and Engineering Research Council and the Alberta Heritage Foundation for Medical Research.

Dr. Elizabeth M. Meiering

Where:	University of Waterloo
Project:	Folding and aggregation of ALS-associated mutant superoxide dismutases
Overview:	Looking at different classes of mutations and then characterising how the mutants behave and how that may be linked to the disease. Has successfully demonstrated a link between SODI mutations and neuronal aggregates.
Funding:	NRP (2003)-Three years (with James R. Lepock)

Dr. Alexander E. Mackenzie

Where:	Children's Hospital of Eastern Ontario (Ottawa)
Project:	Modulation of apoptosis in mouse models of spinal muscular atrophy
Overview:	Examining how cell death can be regulated in spinal muscular atrophy.
Funding:	NRP (2003)-Three years (with Nathalie H. Gendron)

Dr. Stefano Stifani

Where:	McGill University
Project:	Regulation of neuronal development in the mammalian nervous system
Overview:	Examining how the growth and development of neurons is controlled to better understand their function and develop possible treatments for neuromuscular disorders.
Funding:	NRP (2003)-Five years

Dr. Michael J. Ferns

Where:	The Res. Institute of the McGill University Health Centre
Project:	Neurotransmitter receptor localization at the synapse: Regulation by rapsyn
Funding:	NRP (2002)-Three years
Project:	Agrin's role in cholinergic synapse formation
Funding:	NRP (2001) - Three years
Overview:	Understanding how a signaling factor - agrin - directs the formation of the cholinergic synapses. Agrin is required for the formation of the neuromuscular junction.

Dr. Tessa Gordon

Where:	University of Alberta
Project:	A possible link between motoneuronal death and sprouting in ALS
Funding:	Tim Noël Research Award Winner, NRP (2002)-Three years
Overview:	Investigating the effect of injury, exercise and muscle fibre-type in transgenic mice and human tissue to gain insight into the pathways and progress rates for ALS.

Dr. Stephen H. Gee

Where:	University of Ottawa
Project:	The role of diacylglycerol kinase-zeta and syntrophins in neurite outgrowth
Overview:	Studying the molecules that interact with dystrophin to better understand cognitive impairment associated with DMD. Syntrophins link receptors, ion channels, and signaling proteins to dystrophin.
Funding:	NRP (2002)-Three years

Dr. Henry J. Klamut

Where:	University Health Network Ontario Cancer Institute
Project:	Telomerase-mediated lifespan extension applied to the development of autologous myoblast transplantation strategies for DMD
Overview:	Hoping to genetically engineer the patient myoblasts (undeveloped muscle cells) to express dystrophin, or utrophin.
Funding:	NRP (2002)-Three years

Dr. Jeffrey T. Henderson

Where:	University of Toronto
Project:	Role of Eph-B family receptors in regulating motoneuron identity and somatotopic axon outgrowth in the murine spinal cord
Overview:	Studying how neurons communicate with each other during development and following injury. Eph receptors are a family of receptor tyrosine kinases - proteins that regulate communication between cells.
Funding:	NRP (2002)-Three years

Dr. Robin N. Michel

Where:	Concordia University, Montreal
Project:	Calcineurin signaling in the regulation of skeletal muscle fiber growth
Overview:	Michel was the first to demonstrate calcineurin's crucial role in the growth of adult muscle. Working to decipher some of the other factors affecting muscle growth. Calcineurin is found in all muscle types.
Funding:	NRP (2002)-Three years

Dr. David J. Schreyer

Where:	University of Saskatchewan
Project:	Regulation of neuronal phenotype by a muscle-derived factor
Overview:	Investigating the relationship between muscle cells and the motor neurons that target them, with the hope of identifying the factor that affects the way neurons react to injury.
Funding:	NRP (2002)-Three years

Dr. Margaret Fahnestock & Dr. James R. Bain

Where:	McMaster University
Project:	Mechanism of sensory protection of denervated muscle
Overview:	Understand how sensory protection prevents muscle atrophy and determine if sensory protection is a potentially valuable therapeutic approach to nerve damage following injury or disorders such as muscular dystrophies, spinal muscular atrophies and ALS.
Funding:	NRP (2003)-Three years

Dr. Carl-Eric Aubin

Where:	Hôpital Sainte-Justine (Montreal)
Project:	Study of the biomechanical factors related to the progression of scoliotic deformities in DMD
Overview:	Identify the biomechanical factors involved in the progression of spinal curvature in children with DMD and understand the possible pathological pathways of scoliosis.
Funding:	NRP (2001)-Three years

Dr. Salvatore T. Carbonetto

Where:	Montreal General Hospital Research Institute, McGill University
Project:	Dystrophin associated proteins in synapse structure and function
Overview:	Investigating the dystrophin complex in neurons. DMD results from mutations in the gene for dystrophin.
Funding:	NRP (2001)-Three years

Dr. Denis Gravel

Where:	Université de Montreal Institut de réadaptation de Montreal
Project :	Rôle des contractures dans la limitation de la marche des enfants atteints de dystrophie musculaire de Duchenne
Overview:	Studying the biomechanics of how muscle constriction in DMD limits gait
Funding :	NRP (2001)-Three years

Dr. Bernhard Juurlink

Where:	University of Saskatchewan
Project:	Oxidative stress in the CNS and motoneuron disease
Overview:	Investigating how oxidative stress leads to inflammation, and how cellular mechanisms to slow oxidative stress may be enhanced to delay the onset of ALS. Oxidative stress is particularly associated with the familial form of ALS, caused by a mutation in SOD1.
Funding:	NRP (2001) -Three years

Dr. David H. MacLennan

Where:	University of Toronto
Project:	The pathophysiological and genetic basis for muscle diseases resulting from calcium dysregulation
Overview:	Studying how calcium dysregulation can produce muscle disease.
Funding:	NRP (2001)-Five years

Dr. Jean Mathieu

Where:	Centre Hosp. de l'Université de Montreal, Hôpital de Jonquière
Project:	Consequences of neuromuscular genetic disorders: disabilities, social participation, quality of life and their determinants among individuals with myotonic dystrophy
Overview:	Myotonic dystrophy is the most common form of muscular dystrophy in adults. Attempting to measure all factors, whether genetic, personal or environmental, that could play a role in the subject's social participation and quality of life.
Funding:	NRP (2001)-Three years

Dr. Gregory M. Ross

Where:	Queen's University
Project:	Motor neuron death resulting from a zinc-or copper-induced loss of trophic support
Overview:	Conducting basic research to identify critical steps in the nerve cell death process.
Funding:	NRP (2001)-Three years

Dr. Ilona S. Skerjanc

Where:	University of Western Ontario
Project:	The molecular biology of skeletal muscle development
Overview:	Identifying the proteins that are key regulators of muscle development (termed transcription factors) to understand how they function and to determine what controls their ability to function.
Funding:	NRP (2001)-Five years

Dr. Mei Zhen

Where:	Mount Sinai Hospital
Project:	Dissecting molecular mechanisms that regulate the presynaptic differentiation in C. elegans
Overview:	Working on how synaptogenesis is regulated. Neurons establish connections with each other, called synapses, and transmit information from one neuron to another and throughout the body.
Funding:	NRP (2001)-Three years

Dr. Michael J. Strong

Recipient of the Sheila Essey Award (2005)

Where:	Robarts Research Institute (London)
Project:	Intermediate filament expression in sporadic
Funding:	ALS NRP (2001)-Two years
Project:	A molecular and neuropathological characterization of the cognitive impairment of
Funding:	sporadic ALS ALS Canada and The ALS Association (U.S.) - commenced in 2004
Overview:	Discovering the role of neurofilament aggregates in ALS pathways.

Dr. Charles Krieger

Where:	Simon Fraser University
Project:	Functional role of hematogenous inflammatory cells in ALS
Funding:	NRP (2004) - Three years (with Fabio M. Rossi)
Project:	Modulation of neuron death by protein kinase C and calcineurin in ALS
Funding:	NRP (2000)-Two years
Overview:	Attempting to find out why nerve cells die in ALS. In previous work he found that those living with ALS have elevated amounts the enzyme, protein kinase C, which has numerous functions within the cells of the body.

Dr. Jack Puymirat

Where:	Centre Hospitalier, Universitaire de Québec
Project:	Ribozymes and antisense RNA as a tool to study myotonic dystrophy
Funding:	NRP (2000)-Three years, NRP (2003)-Three years
Overview:	Attempting to create gene therapies using "road blocks" that will stop the mutated genes that cause myotonic dystrophy from being read thereby reducing the symptoms of myotonic dystrophy.

Dr. Jacques P.Tremblay

Where:	Université Laval Centre Hospitalier de l'Universté Laval
Project:	Treatment of DMD: Correction of mutated dystrophin mRNA with ribozymes
Funding:	NRP (2001) -Three years
Project:	Autotransplantation of genetically modified myoblasts and muscle derived stem cells
Funding:	NRP (2000)-Three years, NRP (2003) - Three years
Overview:	Growing myoblasts for transplantation in humans.

Dr. John Roder

Where:	Mount Sinai Hospital - Samuel Lunenfield Research Institute
Project:	Novel therapies for ALS
Funding:	NRP (2000)-Three years
Overview:	Exploring the development of effective anti-glutamate agents that may stall or even reverse the relentless progression of ALS.

ADDITIONAL ALS RESEARCHERS

Dr. Shirley Liu

Where:	The Centre for Research in Neurodegenerative Diseases at the University of Toronto
Project:	Involves transgenic mice and the SOD1 gene that is responsible for the hereditary form of ALS. Liu is interested in an active immunization approach to the disease.
Overview:	Provide hope that immunotherapy would have therapeutic potential to help those living with ALS.

Marie Gingras, PhD student

Where:	Currently working with Dr. François Berthod, a researcher and professor at Laval University
Project:	Developing a tissue-engineered model of the spinal cord in ALS research
Overview:	The objective for the project is to develop a three-dimensional model of a reconstructed spinal cord so that the interactions between different cell types and motor neurons can be studied.
Funding:	Holds a studentship from the Fonds de la Recherche en Santé, and funding for the project is provided by a research grant from Muscular Dystrophy Canada.

Glen Hughes, Project Engineer

Where:	Institute of Biomedical Engineering in Fredericton
Project:	Currently working with Dr. Colleen O'Connell, a physiatrist at the Stan Cassidy Centre for Rehabilitation, to develop a proposal for a set of tools that would allow more efficient tracking of the progression of ALS.
Overview:	Eventually such tools could be used for evaluating the effects of different drugs in clinical trials, and to better monitor the diminishing strength of people who have ALS.

Teresa Sanelli, PhD Student

Where:	University of Western Ontario, under the supervision of Dr. Michael Strong
Project:	Working with primary motor neuron cells from transgenic mouse models of ALS that display ALS-like pathology (aggravates) in culture.
Overview:	Examining these processes could give indications as to how to more successfully treat the disease.

Dr. Sanjay Kalra

Where:	University of Alberta
Project:	Using magnetic resonance imaging (MRI) technology to learn more about what could be the cause of ALS and to find biomarkers. Biomarkers - also known as surrogate markers - are biologically-derived indicators that reflect the extent of the disease without having to physically examine tissue.
Overview:	Techniques based on MRI have the potential to improve our understanding of ALS and the way that clinical trials are done.

Dr. Shangxi Xiao

Where:	The Centre for Research in Neurodegenerative Disease at the University of Toronto
Project:	In his research, Xiao's aim is to identify the cause of alternative splicing isoforms, proteins derived from the same gene but with distinct physical and sometimes biological properties; usually a splice variant in the disease.

Dr. Guy Rouleau

Where:	University of Montreal
Project:	Work in his lab has recently led to discovery that the disruption of the peripherin gene in humans accounts for a portion of ALS cases.

François Gros-Louis, PhD Student

Where:	McGill University, with Dr. Guy Rouleau
Project:	Currently screening candidate genes for a study on genetic causes of ALS. He is looking for other genes that may be responsible for the disease or that may affect onset and progression.

Dr. John Turnbull

Where:	McMaster University Hospital
Project:	Researching ways to implement viral delivery vehicles for RNAi (ribo nucleic acid interference). These techniques are being tested in transgenic mice, and also in cell cultures.
Overview:	Turnbull and his team are hopeful that RNAi will have a role in battling familial ALS and, later on, sporadic ALS.

Jason Wilson, PhD Student

Recipient of the Brain Star Award (2005)

Where:	University of British Columbia with Dr. Chris Shaw
Project:	ALS-parkinsonism dementia complex
Overview:	Owing to the overlapping symptoms of several progressive neurological disorders, ALS-PDC may hold the key to unlocking important information about degenerative neurological disorders including ALS.

Dr. Christopher Shaw

Where:	University of British Columbia
Project:	Feeding washed cycad (ancient plant that creates molecules that are toxic) to mouse models to study ALS-parkinsonism dementia complex.
Overview:	With the ALS-PDC models, the disease can be observed in sequence allowing researchers to see what happens before symptoms begin to show.
Funding:	U.S. Army's Defense Department, Natural Sciences and Engineering Research Council of Canada and the Scottish Rite Charitable Foundation of Canada.

Dr. Tim Doherty

Where:	University of Western Ontario
Project:	Improving the ways of measuring disease progression in ALS and other disorders of the motor system.
Overview:	The ability to do so will be crucial to monitor the potential benefit of new treatments as they become available.
Funding:	Compumedics Limited in Australia, the CIHR, the Natural Sciences, Engineering Research Council of Canada (NSERC) and the Canada Research Chairs Program.

Dr. Neil Cashman

Where:	The Centre for Neurodegenerative Diseases, University of Toronto Direct - The ALS Centre at G.F. Strong Rehabilitation Centre (B.C.) and the University of British Columbia, Canada Research Chair in Neurodegeneration and Protein Misfolding Diseases (which includes ALS along with Prion diseases, Alzheimer disease and Parkinson"s disease) - January 2006
Overview:	Laboratory research focuses upon the cellular and molecular basis of neurodegenerative diseases.