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## Research News

JULY 2007 - VOLUME 15

Welcome to Research News. This newsletter is sent to those who have signed up for ALS Society of Canada bulletins, the members of the ALS Society of Canada board of directors, provincial society staff, ALS researchers, ALS unit board members, ALS clinics, ALS society volunteers, and international ALS/MND organizations. If you wish others to receive this newsletter, please forward e-mail addresses to Bobbi Greenberg – [bg@als.ca](mailto:bg@als.ca) – requesting inclusion in the UPDATE e-list.

*In this newsletter we are bringing together and reporting on current research. ALS Canada does not assume responsibility for the information contained in this newsletter.*

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### NEWS FROM AMERICAN ACADEMY OF NEUROLOGY ANNUAL MEETING

**T**he results of a nine-month trial of minocycline for ALS were announced at the American Academy of Neurology Annual Meeting in Boston, held April 28 - May 5. The trial tested minocycline vs a placebo in 412 people at 31 American centres. **Minocycline demonstrated no beneficial effect, and for some patients, worsened measurable outcomes.** In the trial, the drug did not affect survival or quality of life measures for people with ALS.

A 20-week trial of sodium phenylbutyrate in 40 people with ALS at eight centres

showed the drug was safe and tolerable for the majority of participants at dosages between 12 and 21 grams per day. There were no significant abnormalities noted in blood tests, weight or vital signs of those in the study. The next step, pending regulatory approvals, will be an efficacy study. Sodium phenylbutyrate is a marketed drug and prescribed for children with certain metabolic abnormalities. The U.S. Food and Drug Administration has requested animal safety studies before giving regulatory approval for tests of drug efficacy in patients.

### A RANDOMIZED CONTROLLED TRIAL OF RESISTANCE EXERCISE IN INDIVIDUALS WITH ALS

**T**he objective of this study (conducted by Vanina Dal Bello-Haas, PT, PhD, at the University of Saskatchewan and her American colleagues) published in the June 5 issue of *Neurology* was to determine the effects of resistance exercise on function, fatigue, and quality of life in individuals

with ALS. The study found those who did moderate strengthening exercises had a 12 per cent slower decrease in function and a 16 per cent slower decline in quality of life over the six months than those who did the stretching exercises alone. For more information [click here](#).

## FOCALITY OF UPPER AND LOWER MOTOR NEURON DEGENERATION AT THE CLINICAL ONSET OF ALS AND IMPLICATIONS OF ALS FOCALITY

In these two studies (published in *Neurology*) Dr. John Ravits and colleagues at Virginia Mason Medical Center in Seattle, Washington, explore the body region of first symptoms of ALS in 100 patients, then assess the nature of lower motor neuron loss in 19 ALS post-mortem nervous systems. In 98 per cent of patients, ALS began in one body region with upper and lower motor neurons signs being maximal at this same body region. The spread of upper motor neuron (UMN) and lower motor neuron (LMN) signs occurred in a sequential process from both original UMN and LMN foci more often in the direction of more caudal body regions.

In the second study, the actual loss of LMNs from the original focus was graded. In the majority of post-mortem samples, motor neuron loss proceeded in a radial pattern of degeneration. These studies emphasize the anatomical basis of disease progression in ALS and may allow for more precise determination of anticipated location of subsequent clinical signs, as well as identification of nervous system regions which might profit from specific targeting to save or rescue motor neurons if a regionalized therapeutic strategy is designed. For more information [click here](#) (UMN and LMN study) or [click here](#) (focality study)

## PROGNOSIS OF ALS RESPIRATORY ONSET

Dr. Christen Shoesmith and colleagues at the Department of Clinical Neurological Sciences, University of Western Ontario published this paper in the *Journal of Neurology, Neurosurgery, and Psychiatry*.

The authors discussed respiratory muscle involvement in ALS. They reported on the clinical features and prognosis of 21 patients with respiratory onset ALS. Non-invasive positive pressure ventilation (NIPPV) significantly improved survival compared with those who did not use NIPPV. The study suggests that ALS with respiratory onset does not necessarily follow a rapidly progressive course.

## RESEARCHERS HAVE NEW INSIGHT INTO ALS

University of Kentucky researchers have discovered a new cellular mechanism that may better explain what causes ALS. In a paper published in the June 1 issue of the *Journal of Biological Chemistry*, UK molecular and cellular biochemistry assistant professor Haining Zhu reported on how mutations in the gene that makes SOD1 slows down the intracellular transport of molecules in

neurons. The results of the study by Zhu and his colleagues show that the defective transport of molecules is probably due to the aberrant interaction between the disease-causing SOD1 mutants and a motor complex that is essential to the intracellular transport in neurons. This provides new insight into ways to prevent or slow ALS. For more information [click here](#).

## SAVE THESE DATES

### 18TH INTERNATIONAL SYMPOSIUM ON

**ALS/MND** - The meeting will take place December 1-3 in Toronto. The ALS Society of Canada is the host this year. The symposium brings together leading international researchers and health-care professionals to present and debate key innovations in their respective fields.

**CLINICAL MEETING** - Chairs Dr. Michael Strong, director of ALS Centre at the University of Western Ontario and Dr. Wendy Johnston, co-director of the ALS clinic at the University of Alberta will host this session, which will explore new developments in diagnosis and management of ALS/MND and the relationship between clinical experience, research methodologies and influencing activities in enhancing quality of care, now and in the future.

**SCIENTIFIC MEETING** - Chair Jean-Pierre Julien, PhD, Canada Research Chair in the Mechanisms of Neurodegeneration at Laval University, will be hosting this meeting, which aims to combine established theories on motor neuron function, dysfunction and degeneration, across the broad spectrum of motor neuron disease, with emerging avenues of therapeutic investigation.

For more information [click here](#).