

ALS Society of (Windsor) Essex County Endowment in Support of Students in ALS Research

Research Report 2019: Dr. Michael Strong

It would not be an overstatement to say that the support of the ALS Society of Windsor-Essex County continues to be transformative in helping to advance our understanding of the basic mechanism(s) of ALS. Your support has been critical in three key areas:

- 1. Development of a novel fly model of ALS;
- 2. Providing critical support to summer studentships; and
- 3. Providing much needed support for equipment that cannot be achieved through any other route.

In this summary, we will provide an update in each of these areas.

Development of a new model of ALS in a fly

In our last update, we had started a fly project looking at characterizing the expression of a new protein that appears to be critical in controlling key elements of RNA metabolism that are disrupted in ALS. This protein is call RGNEF. This work has been solely funded by the ALS Society of Windsor-Essex and in doing so allowed us to develop the critical core data to prove that RGNEF can profoundly alter the course of neurodegeneration in a fly that has been genetically modified to have many of the features of ALS. Further, and has been the case with much of the support from your group over the last three decades, it provided us with the much needed data to further the experiments by obtaining long term funding.

Taking care of the caregivers

The Schulich School of Medicine & Dentistry family are defined and supported by four pillars – altruism, responsibility, respect and integrity. These values, combined with the support provided by donors, ensure our school has long been recognized as graduating health professionals who are compassionate and meet the highest standards of care.

Together, we help take care of the caregivers, whether that is supporting students financially, finding new ways to be creative and promote learning, and even case-based learning where students work in small teams to resolve real-life situations.



Together, we are finding answers to research questions

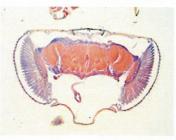
Providing critical support to summer studentships

In each of the last few years, we have been tremendously fortunate to use the summer student support provided by the ALS Society of Windsor-Essex to bring in talented students who then proceed to complete a graduate degree with us. This provides a critical piece in the puzzle of training the next generation of ALS researchers. Recall that past student, Michael Tavolieri, was an incredible addition to our lab – his summer studentship allowed him to get a head start on his research project. You will be pleased to know that his manuscript derived from this work was published this past January and is already being critically cited by others in the field. He also successfully defended his thesis.

This same approach was used in recruiting Benjamin Withers to our lab; first a summer position supported by the society and then a MSc program from which Ben graduated this January. Ben was integrally involved in developing the fly model described above and specifically examining how RGNEF actually works. It has meant doing two genetic manipulations of the flies, a manipulation that when present leads to red eyes and wrinkled wings as markers so that we can tell which flies have the ALS characteristics (see top photo on right). We can then take the flies and look at their brains (see lower photo on right) in order to see if RGNEF has altered the disease process. We are very excited to let you know that in fact, RGNEF looks to be able to substantially reduce the burden of disease in this model, in some cases, actually preventing it from occurring!



The Schulich School of Medicine & Dentistry also has a program that provides medical students the opportunity to conduct research during the summers. We have partnered with this program using the summer student funding to support Alex Martin, who is a medical student from the Windsor Campus of the Schulich School. Alex completed his first summer last year and is returning this summer to complete his study. He is an incredibly gifted young individual whose research project is examining how the cells that are profoundly



damaged in ALS differ from those that are completely spared even though both populations of cells are motor neurons. To do this, we are collaborating with Dr. Robert Hammond who is a neuropathologist to identify these uniquely spared cells. It is a study that has never been done before and can only be done because of the unique research of our lab into the fundamental molecular mechanisms of ALS. It is our sincere hope that Alex will become a future clinician scientist with a focus on ALS.

The summer studentship also supported the entry of Matthew Hintermayer prior to his MSc with us; he is preparing to defend his thesis this summer and will then commence a MD/PhD program at McGill University in Montreal. Matt has been studying an unusual form of ALS that occurs in athletes and which we have demonstrated can be induced in rats following a head injury. This work was published last year; Matt is following up to define the mechanism. The importance is that both ALS as we normally see it, and that which can occur in athletes with head injuries, looks to have similar mechanisms of disease process. Understanding one will have critical implications for the other, and in particular the dementia of ALS. It is no small coincidence that the whole field of understanding the dementia of ALS is one which was pioneered in our lab and which would never have happened without your support. It is thus fitting that a student supported initially by your summer student program should provide the answer as to how this happens. It goes without saying that we are exceptionally proud of all of these students. The ALS Society of Windsor-Essex should equally and justifiably be proud to have supported them.

Together, we have made Western stronger

Providing much needed support for equipment

From the first support of our research by providing a computer so that we could track our ALS patients to this day, the support of the ALS Society of Windsor-Essex has been critical to our success. This is not always glamorous, but it is critical since there are no mechanisms to gain support of this nature at any level of peer-reviewed support. This past year, your support allowed us to make three critical purchases. Firstly, and as part of the development of the fly model, we required a new centrifuge. Because of advancements in biosafety regulations we require biocontainment equipment, and funds provided by the society allowed us to purchase a state-of-the-art biosafety containment centrifuge that meets the regulations for our work (pictured at right). We also replaced some standard equipment that we use every day, including battery powered manual pipettors, vortexes and gel electrophoresis equipment. With the large number of people working in the lab this allows us to not have to wait for equipment availability or to borrow equipment from other labs to conduct our work.



We are also tremendously grateful for the support for the purchase of the SPR unit. This unit is only one of two on the University campus, with the other being a teaching unit that is not accessible. It arrived in our lab in late January. A technician has been hired specifically to oversee its operation (Crystal McLellan, PhD). This will add a truly unique dimension to our research as we delve further into the fundamental basis of ALS. In the next year, we are very much looking forward to demonstrating to you how this has advanced the field of RNA biology in ALS.

Thank you

As you are aware, I have taken on the job of President of the Canadian Institutes of Health Research and had to close my clinical practice. This was not an easy decision as I have been grateful to be able to help so many patients and their families over the years as they undertook this difficult journey with ALS. However, I have not left the research behind, and indeed, our lab is larger and busier than ever. We have secure funding for our research program for the next five years; funding that could not have been obtained without all of the generosity and support of the ALS Society of Windsor-Essex County. Whether it be direct support of new projects, student support, or critical equipment, you have been crucial to our success. For this we are grateful.

I look forward to providing you a report in 2020 that details even further how this support continues to be transformative.

With best regards, Dr. Michael J. Strong

