Fondation de l'Ataxie
Charlevoix-Saguenay

OFFER FOR FINANCING RESEARCH ON ARSACS

1. **Goal:** The Ataxia of Charlevoix-Saguenay Foundation offers annual research fellowships that will lead to a treatment for autosomal recessive spastic ataxia of Charlevoix-Saguenay (ARSACS). See Appendix I attached.

2. **Required documents:**
   - Completed application form.
   - Description of the research project
   - CV of the Applicant.

3. **Amount:** A maximum of $100,000 could be awarded for a period of one year and could be renewed for a second year by way of a new application. **Also, for this year, a funding application for a specific project could include several labs in Canada or elsewhere. In such a case, the $100,000 limit would not apply.**

4. **Number of scholarships awarded each year:** To be determined.

5. **Competition Deadline:** May 25, 2018.

6. **Submission:** Applicants must e-mail the completed form (including annexes) at the latest the day of the competition deadline to Sonia Gobeil at the following address: sgobeil@ctf.ca

7. **Bi-Annual Report:** The Ataxia of Charlevoix-Saguenay Foundation will request a bi-annual written report from the candidate in the project being supported by the Foundation. Such a report, which will be sent directly to the Foundation, will state the scientific achievements and scientific goals of the project along with the expenses incurred and projected by the candidate.

8. **Evaluations:** The Scientific Advisory Board of the Ataxia of Charlevoix-Saguenay Foundation will evaluate each application.

Fondation de l'Ataxie
Charlevoix-Saguenay

FINANCING OFFER FOR RESEARCH ON ARSACS

APPLICATION FORM

IDENTIFICATION OF APPLICANT	
SURNAME :	**FIRST NAME :**
ADDRESS :	**TEL. WORK :** **FAX :** **EMAIL :**
DATE OF BIRTH (Year/Month /Day)	**CITIZENSHIP :** ☐ Canadian ☐ Permanent Resident of Quebec ☐ Other : _____
LANGUAGE OF CORRESPONDENCE ☐ French ☐ English	
TITLE OF THE RESEARCH PROJECT ON ARSACS :	

APPLICANT'S DECLARATION

I declare that the information provided is true an accurate and agree to the conditions described in this form for the financing opportunity.

_____ _____

Signature Date

DOCUMENTS TO PROVIDE

The Applicant must submit the following documents by email in PDF format to the attention of Sonia Gobeil: sgobeil@ctf.ca

1. **APPLICATION FORM :** The applicant form duly completed.

2. **RESEARCH PROJECT :** The Applicant must describe the research project for which funding if requested (maximum 3 pages).

3. **APPLICANT'S CV :** Please provide a short CV.

APPENDIX I

ARSACS is the common name for Autosomal Recessive Spastic Ataxia of Charlevoix-Saguenay. This condition was first seen in people of the Charlevoix-Saguenay region of Quebec, Canada. The majority of people with ARSACS live in Quebec or have recent ancestors from Quebec. People with ARSACS have also been identified in various countries such as Japan, Turkey, Tunisia, Spain, Brazil, Poland, Italy, and Belgium. It is a progressive disease that affects the body's ability to create a protein called sacsin, normally found in the brain, skin, and muscles. Over 170 SACS mutations have been reported worldwide and are thought to cause loss of function of sacsin, a poorly characterized and massive (520 kDa) protein.

Research suggests that sacsin might play a role in folding newly produced proteins into the proper 3-dimensional shape because it shares similar regions with other proteins that perform this function. Mutations in the *SACS* gene cause the production of an unstable sacsin protein that does not function normally. It is still unclear how the abnormal sacsin protein affects the brain and skeletal muscles and results in the signs and symptoms of ARSACS.

In existing in vitro (patients' cell lines) and in vivo (Sacs(-/-) mouse) models, a disruption of mitochondrial transport is observed along with abnormal accumulation of non-phosphorylated neurofilament (NF) bundles in the somatodendritic regions of vulnerable neuronal populations, a significant reduction in mitochondrial motility and elongated mitochondria. The data points to alterations in the NF cytoskeleton and defects in mitochondrial dynamics as the underlying pathophysiological basis of ARSACS.

There are currently no treatments available to cure people with ARSACS.

The Ataxia Charlevoix-Saguenay Foundation, funded in 2006, is a charitable organization federally registered with no employees and is supported entirely by private donations and volunteers to fund scientific research on ARSACS. The Ataxia Charlevoix-Saguenay Foundation's mission is to discover and develop a treatment for the disease.

The present call for proposals aims to funds projects that will clearly advance the understanding of the disease and lead the way to the development of a treatment for ARSACS patients.