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Research supported by the Huntington Society of Canada (HSC) provides new insight into Huntington disease (HD) and may provide benefits for other neurodegenerative diseases

(KITCHENER, ON) July 10, 2018 – A multi-institutional, international study supported in collaboration by the Huntington Society of Canada (HSC), the Krembil Foundation, the Canadian Institutes of Health Research (CIHR) and McMaster University has developed a new theory on Huntington disease (HD) which is showing promise and is opening new avenues of potential drug development.

HD is a debilitating brain disorder that is fatal and incurable. About one in every 7,000 Canadians has HD and approximately one in every 5,500 is at-risk of developing the disease.

HD is caused by a mutation in the gene that makes the protein called huntingtin. This study was conducted in partnership with the Ray Truant lab of McMaster University, the Simonetta Sipione lab of the University of Alberta, the David Litchfield lab of the Western University and the Christopher Ross Lab of Johns Hopkins University. They found that there is a unique type of signalling coming from damaged DNA, that signals huntingtin activity in DNA repair, and that this signalling is defective in Huntington disease.

A study developing the new hypothesis was published this week in the *Proceedings of the National Academy of Science (PNAS)*.

"The concept was that if we applied the signalling molecule back in excess, even orally, this signalling can be restored in the Huntington disease mouse brain," said Laura Bowie, a PhD student in the Department of Biochemistry and Biomedical Sciences at McMaster. "This resulted in the mutant huntingtin protein levels being restored to normal, leading to a correction of Huntington disease in mouse models."

This discovery was made using a robotic microscope and drug screening by artificial intelligence, coupled to state-of-the-art super-resolution microscopy in a system developed by Bowie.

Ray Truant, senior author on the study, has focussed his career on Huntington disease research and how the mutation leads to Huntington disease. His lab was the first to show that normal huntingtin was involved in DNA repair.

"This is an important new lead and a new hypothesis, but it is important for people to know this is not a drug or cure," said Truant, professor in the Department of Biochemistry and Biomedical Sciences at McMaster. "These compounds also show promise in Parkinson disease models as there may be pathways in common for Huntington and Parkinson diseases."

Dr. Truant's research is supported in part through HSC's research funding strategy which supports excellent, peer reviewed research into the most promising basic and clinical research, leading to viable treatments for



HD. The universal goal for the international HD research focus is to find treatments that reverse, slow or prevent the progression of HD.

"The Huntington Society of Canada is proud to support such leading edge research," says Bev Heim-Myers, CEO of the Huntington Society of Canada. "Innovative research initiatives, such as the work led by the team in Dr. Truant's Lab, including PhD student Laurie Bowie, has the potential to transform HD research. The answers we find for Huntington disease will likely lead to better understanding of treatments for other neurological diseases and it is important that we continue this cross-talk amongst neurodegenerative diseases."

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Huntington disease (HD) is a debilitating brain disorder that is fatal and incurable. About one in every 7,000 Canadians has HD and approximately one in every 5,500 is at-risk of developing the disease. Many more are touched by HD whether as a caregiver, a family member, or a friend. HD causes cells in specific parts of the brain to die. As the disease progresses, a person with Huntington disease becomes less able to manage movements, recall events, make decisions and control emotions. Many describe the symptoms of HD as having ALS, Parkinson's and Alzheimer's – simultaneously.

The **Huntington Society of Canada (HSC)** is a respected leader in the worldwide effort to end Huntington disease. HSC is the only Canadian health charity dedicated to providing help and hope for families dealing with Huntington disease across Canada.

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