HEIDELBERG, 20 January 2014 – Scientists from the **Montreal Neurological Institute** and Hospital in Canada have discovered that
two genes linked to hereditary Parkinson's disease
are involved in the
early-stage quality control of mitochondria
. The protective mechanism, which is reported in
The EMBO Journal
, removes damaged proteins that arise from
oxidative stress from mitochondria
.

"*PINK1* and *parkin*, are implicated in selectively targeting dysfunctional components of mitochondria to the lysosome under conditions of excessive oxidative damage within the organelle," said Edward Fon, Professor at the McGill Parkinson Program at the Montreal Neurological Institute and Hospital. "Our study reveals a quality control mechanism where vesicles bud off from mitochondria and proceed to the lysosome for degradation. This method is distinct from the degradation pathway for damaged whole mitochondria which has been known for some time. It is also an early response, proceeding on a timescale of hours instead of days."

The deterioration of mechanisms designed to maintain the integrity and function of mitochondria throughout the lifetime of a cell has been suggested to underlie the progression of several neurodegenerative diseases, including Parkinson's disease. When mitochondria, the "power plants" of the cell that provide energy, malfunction they can contribute to Parkinson's disease. If they are to survive and function mitochondria need to degrade oxidized and damaged proteins.

In the study, immunofluorescence and confocal microscopy were used to observe how the vesicles “pinch off” from mitochondria with their damaged cargo. “Our conclusion is that the loss of this *PINK1* and *parkin*-dependent trafficking system impairs the ability of mitochondria to selectively degrade oxidized and damaged proteins and leads, over time, to the mitochondrial dysfunction noted in hereditary Parkinson’s disease,” said Heidi McBride, Professor in the Neuromuscular Group in the Department of Neurology and Neurosurgery at the Montreal Neurological Institute and Hospital.

Both salvage pathways are operational in the cell. If the vesicular pathway, the first line of defense, is overwhelmed and the damage is irreversible then the entire organelle is targeted for degradation.

Parkin and PINK1 function in a vesicular trafficking pathway regulating mitochondrial quality control

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