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## Media Release

From the Minister of Innovation

May 07, 2007 09:00 ET

### Melbourne Researchers Uncover Underlying Cause of Mitochondrial Diseases Findings to Be Presented at BIO 2007, Published in Molecular Biology of the Cell

MELBOURNE, AUSTRALIA and BOSTON, MA -- (MARKET WIRE) -- May 7, 2007 -- Mitochondrial diseases, a class of rare and incurable conditions, are believed to result from the failure of mitochondria in the cells to produce a universal energy carrying molecule called ATP (adenosine triphosphate). Now research conducted at Melbourne's LaTrobe University suggests that a signaling problem in the cells is at fault, turning the commonly held theory on its head. The findings have implications for development of drug therapies to treat the many forms of mitochondrial disease as well as for most major neurodegenerative disorders, where mitochondrial dysfunction has been demonstrated to play a central role. These include Amyotrophic Lateral Sclerosis (Lou Gehrig's Disease), Parkinson's, Huntington's and Alzheimer's disease. The research will be presented at the BIO2007 conference in a poster to be located in the Innovation Corridor, and a publication is planned in the journal Molecular Biology of the Cell.

"This research gives us a completely novel understanding of the causal mechanisms of mitochondrial disease, demonstrating that such conditions may result from a signaling disorder in the cells, rather than a fundamental energy insufficiency as was previously thought," said Paul Fisher, Ph.D., lead investigator and Chair in Microbiology at LaTrobe University.

"We are hopeful that this finding will lead to effective new approaches to treating both rare and prevalent diseases involving mitochondrial dysfunction," said John Brumby, Minister of Innovation, Victoria, Australia.

"Like a smoke alarm that activates at the first sign of trouble, AMPK is an energy-sensing alarm protein that averts an impending energy crisis in the cell by activating before the situation becomes critical," said Dr. Fisher. "When triggered, AMPK temporarily shuts down a variety of activities and initiates energy production within the cell. In a healthy cell this returns ATP supplies to normal and allows the cell to return to regular functioning.

"What we found is that in a mitochondrially diseased cell, the AMPK alarm is permanently activated," Dr. Fisher said. "Many of the cellular outcomes of mitochondrial dysfunction are known also to be associated with AMPK signaling, but this is the first time the 'alarm protein' has itself been implicated in mitochondrial disease causation."

#### About the Research

Using the slime mould *Dictyostelium discoideum* as a model for mitochondrial disease, researchers genetically manipulated the signaling pathways that control cellular functions thought to be involved in mitochondrial disease. Overproduction of an active form of AMPK was shown to create the same symptoms as mitochondrial disease, while decreasing the supply of AMPK completely suppressed those symptoms in "Dicty."

#### About Mitochondrial Disease

About one in 4,000 children in the United States will develop mitochondrial disease by the age of ten. Any organ or tissue can be affected by mitochondrial disease, but it is generally the central nervous system, muscles, hearts and less often the kidneys or insulin-producing cells of the pancreas that are affected. Overtly mitochondrial diseases are rare but incurable and range in severity from mild to fatal. Friedreich's Ataxia, MERRF (Myoclonic Epilepsy and Ragged Red Fibres and MELAS (Myoclonic Epilepsy, Lactic Acidosis and Stroke-like Episodes) are examples of mitochondrial diseases.

The World Health Organization (WHO) calculates that neurodegenerative diseases, also associated with mitochondrial dysfunction, will become the world's second leading cause of death by the year 2040.

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