Molecular mechanisms of neurodegeneration caused by mitochondrial disfunction

https://www.neurodegenerationresearch.eu/survey/molecular-mechanisms-of-neurodegeneration-caused-by-mitochondrial-disfunction/

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Country

Finland

Title of project/programme

Molecular mechanisms of neurodegeneration caused by mitochondrial disfunction

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Academy of Finland

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The project/programme is most relevant to:

Neurodegenerative disease in general

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Neurodegeneration | mitochondria | fibroblast growth factor 21 | mitochondrial DNA polymerase gamma | single-stranded DNA-binding proteins | tumor suppressor p53 | mitochondrial recessive ataxia syndrome | infantile onset spinocerebellar ataxia

Research Abstract

This study focuses on two severe mitochondrial neurodegenerative disorders – mitochondrial recessive ataxia syndrome, MIRAS, and infantile onset spinnocerebellar ataxia (IOSCA). These diseases are the most common ataxias in Finland. MIRAS is caused by recessive mutation in mitochondrial DNA polymerase gamma (POLG), while IOSCA by recessive mutation in mitochondrial helicase Twinkle. Symptoms of these diseases are very similar except that IOSCA manifests very early in life starting from 9-18 months after birth. The reasons why these mutations cause such severe phenotypes is unclear and this study seeks to answer this question. To date, no effective treatments exist for mitochondrial neurodegeneration. The second aim of this study is to use employ endocrine pathway for physiological regulation of mitochondrial neurodegeneration. This multidisciplinary study recruits a variable set of tools involving protein chemistry, animal models for MIRAS and IOSCA and disease-relevant cell cultures.

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Fellowships

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