Molecular neuropathology of TDP-43 proteinopathies

https://neurodegenerationresearch.eu/survey/molecular-neuropathology-of-tdp-43-proteinopathies-2/

Principal Investigators

LEE, EDWARD BYUNG-HA

Institution

UNIVERSITY OF PENNSYLVANIA

Contact information of lead PI
Country

USA

Title of project or programme

Molecular neuropathology of TDP-43 proteinopathies

Source of funding information

NIH (NINDS)

Total sum awarded (Euro)

387500

Start date of award

15/05/2016

Total duration of award in years

2

Keywords

protein TDP-43, frontotemporal degeneration, neuropathology, Amyotrophic Lateral Sclerosis, Nuclear RNA

Research Abstract

Project Summary Amyotrophic lateral sclerosis (ALS) and frontotemporal degeneration (FTD) are two related neurodegenerative diseases which share overlapping clinical, pathologic and genetic features. The ALS-FTD spectrum of diseases is uniformly fatal, and there is neither treatment nor cure. There is a remarkable convergence of pathologic and genetic data which indicate that abnormal RNA metabolism is linked to neurodegeneration in ALS and FTD. The

overall goal of this proposal is define the molecular neuropathology of ALS and FTD using human brain tissue. Understanding the molecular aberrations associated with specific brain pathologies will reveal insights into the molecular pathogenesis of ALS/FTD. Novel methods are presented in which pathologic proteins or neurons are isolated from human brain for deep molecular analysis. Two specific aims are proposed which will (1) determine the molecular composition of pathologic TDP-43 protein inclusions, and (2) identify the abnormalities in RNA processing due to the loss of normal nuclear TDP- 43 protein. These studies will further our understanding of TDP-43 proteinopathies by using highly innovative techniques to study human brain tissue with advanced molecular techniques.

Further information available at:

Types:
Investments < €500k

Member States:
United States of America

Diseases:
N/A

Years:
2016

Database Categories:
N/A

Database Tags:
N/A