

From test-tubes, to mice, to human ALS

<https://neurodegenerationresearch.eu/survey/from-test-tubes-to-mice-to-human-als/>

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Country

Sweden

Title of project or programme

From test-tubes, to mice, to human ALS

Source of funding information

The Swedish Brain Foundation

Total sum awarded (Euro)

€ 108,814

Start date of award

01/07/2015

Total duration of award in years

2.5

Keywords

Research Abstract

Despite decades of intense research, there is yet not a single spinal-cord sample from a presymptomatic or in-progression ALS patient: all existing information is from post-mortem autopsy of patients with fully advanced disease and this has proved insufficient for establishing why and how it all starts. As a consequence, mechanistic studies of ALS are currently limited to biophysical studies in vitro, cell cultures, and transgenic mice, where the latter is the only tractable tissue-level model for the human disease. With this in hand, our research has become focused on establishing – at molecular level – what is going on in the transgenic ALS mice, and how these pathological events are mechanistically linked to the physical-chemical properties and aggregation behaviour of the causative protein SOD1. Although this could seem as a straightforward proposition, it relies critically on solving two problems that has so far eluded the scientific community. First, to find how out the protein behaviour in vitro translates to the much

more complex conditions in live cells. Second, to obtain atomic-level structural and quantitative information about the SOD1 molecule in intact neural tissue that is not amenable to analysis by conventional biophysical methods

Further information available at:

Types:

Investments < €500k

Member States:

Sweden

Diseases:

N/A

Years:

2016

Database Categories:

N/A

Database Tags:

N/A