Neurodegeneration

Principal Investigators

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Institution

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Contact information of lead PI
Country

United Kingdom

Title of project or programme

Neurodegeneration

Source of funding information

MRC

Total sum awarded (Euro)

€ 2,116,735

Start date of award

01/01/2012

Total duration of award in years

5.0

The project/programme is most relevant to:

Huntington's disease|Motor neurone diseases

Keywords
Research Abstract

Our major aim is to identify and elaborate new genetic pathways involved in neurodegeneration using disease mouse models. In particular, we focus our work on two major neurodegenerative disorders: Huntington's disease (HD) and amyotrophic lateral sclerosis (ALS). Despite intense research focus on HD and ALS, they both remain incurable, and the only available treatment licensed for ALS, Riluzole, has limited therapeutic benefits. Although both disorders are very different in their genetics, they are both fatal and exemplify the need for a deeper understanding of disease pathogenesis as the base on which to build the search for therapeutic intervention.

Mouse models are critical in our current understanding of disease processes, including neurodegeneration. The focus of our programme is to elaborate on disease pathogenesis by studying neurodegeneration mouse models. We work with both established neurodegeneration mouse models as well as generating our own new mouse models, making them immediately available to the research community. We uncover new genes and mechanisms leading to neurodegeneration by means of two major methodologies: First, the generation of new disease mouse models carrying point mutations in known neurodegenerative causative genes, their behavioural characterisation and the subsequent elucidation of the molecular mechanisms leading to disease. We are currently focussing on ALS genes using this approach, and have uncovered new ALS mouse models carrying point mutations in Sod1 and Tardbp (the gene encoding TDP43). Second, the identification of new genes involved in neurodegeneration by means of ENU mutagenesis forward genetics screening. The screening pipelines used include modifier screens over well established disease mouse models (both HD and ALS models) as well as the identification of new neurodegenerative genes through the Harwell ENU ageing screen pipeline. Our genetic work is set in the context of our key links into other leading UK laboratories bringing genetic, biochemical and cell biological studies to bear. The new mouse models provided, together with the new disease pathways identified would be valuable tools in the further elucidation of neurodegeneration pathophysiology by us and others.

Lay Summary
Further information available at:

Types:
Investments > €500k

Member States:
United Kingdom

Diseases:
Huntington's disease, Motor neurone diseases

Years:
2016

Database Categories:
N/A

Database Tags:
N/A