Transgenic modelling of human prion diseases, intermammalian transmission barriers & assessing candidate therapeutics

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Principal Investigators

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Contact information of lead PI
Country

United Kingdom

Title of project or programme

Transgenic modelling of human prion diseases, intermammalian transmission barriers & assessing candidate therapeutics

Source of funding information

MRC

Total sum awarded (Euro)

€ 8,813,360

Start date of award

01/04/2011

Total duration of award in years

5.0

The project/programme is most relevant to:

Prion disease

Keywords
Research Abstract

In studies now spanning well over a decade we have developed highly specialised transgenic transmission facilities for comprehensive studies of human prion disease under appropriate

biocontainment. Species-barrier-free transmission of many forms of human prion disease is available. While excellent, well characterised models are now in routine use, further improvement to these models, and the development of improved models for the assay of vCJD prions are underway. In addition to ongoing basic research into intermammalian and strain-specific transmission barriers and characterisation of the prion strains causing human disease, this facility is being applied to address key public health issues which require human prion bioassay (tissue distribution of infectivity and human prion decontamination for example) and to study putative prion therapeutics. Animal models of a range of inherited human prion diseases are being studied, in particular to determine if prions are produced spontaneously in such animals. The specific aims of this programme are as follows:- To establish the full range of prion strains causing human disease and to attempt to biologically clone these for biochemical study (with programme 6) – To complete our long-term characterisation of intermammalian prion transmission barriers of relevance to public health and to understand the interaction between prion strain diversity and these barriers – To model the human inherited prion diseases and determine whether prions can be generated spontaneously in these models – To generate new transgenic lines as required to model novel human PrP polymorphisms and candidate prion modifier genes (in conjunction with programmes 1 and 2) – To generate transgenic models of frontotemporal dementia (in conjunction with programme 10) – Development and use of standardised models for evaluation of candidate therapeutics – To continue to provide and update the Units Transgenic Core facility and support animal research throughout the Unit

Lay Summary
Further information available at:

Types:
Investments > €500k

Member States:
United Kingdom

Diseases:
Prion disease

Years:
2016

Database Categories:
N/A

Database Tags:
N/A