

Strain Typing of vCJD Cases

<https://neurodegenerationresearch.eu/survey/strain-typing-of-vcjd-cases/>

Principal Investigators

Professor Jean Manson

Institution

The Roslin Institute

Contact information of lead PI

Country

United Kingdom

Title of project or programme

Strain Typing of vCJD Cases

Source of funding information

NIHR (PRP Ad Hoc Grants)

Total sum awarded (Euro)

€ 1,950,535

Start date of award

01/08/2011

Total duration of award in years

5.5

The project/programme is most relevant to:

Prion disease

Keywords

Research Abstract

Background:

vCJD was first reported in the UK in 1996 by NCJDSU. Since then, 171 deaths from probable or definite vCJD have occurred, including 3 in blood transfusion related cases. A case of asymptomatic vCJD infection following blood transfusion was also reported. vCJD has not just been restricted to the UK; 49 cases have been reported in 10 other countries. Initial studies have demonstrated strong similarities between strains from different countries but differences in incubation time have been observed which will be examined by sub-passage. This will confirm

whether cases from non-UK countries and the UK are of the same strain and to show if any strain differences have occurred over time in the UK and other countries.

Aims:

In collaboration with the National CJD Surveillance Unit we propose to use our well-defined strain typing panels of wild type and humanised transgenic mice to characterise vCJD cases of non-UK (French and USA) and UK origin (both recent, atypical and historical) of vCJD. By doing so we aim to answer:

- whether non-UK and UK cases of vCJD are of the same disease strain?
- has any strain modification occurred between early and late occurring cases of vCJD in the UK?
- investigate an atypical case of vCJD in an elderly person: has a new strain emerged or has the patient's age and confounding factors impacted on the clinical features of disease?

Plan of Investigation:

Eight cases of vCJD; 2 non UK (French and USA), 4 recent (the donors and recipients of blood transfusion related cases) and the brain and spleen of a historical case will undergo a secondary passage into a strain typing panel of mice. Two historical cases of vCJD, 2 donors to blood transfusion cases and a more recent case will undergo a primary transmission into a similar panel. A primary transmission of a vCJD case exhibiting atypical clinical disease characteristics will also be inoculated into panels of wild type and humanised transgenic mice in order to assess transmission potential. Transmission studies will be set up over years 1 and 2, and over years 2, 3 and 4 of the proposal, pathological and biochemical analysis and comparison of the strain properties will be carried out.

Potential Impact:

By identifying potential new strains of disease or changes in the pattern of disease we increase our understanding of the infectious agent, this will impact on both diagnosis of disease and aid in CJD surveillance programmes. This proposal will identify whether cases of vCJD from different countries are of the same strain and whether there have been any strain modifications between cases diagnosed at the beginning or the latter stages of the current epidemic. We are also investigating a case of vCJD in an individual whose clinical onset was significantly later than the mean and whom exhibited atypical clinical symptoms. This may be a new strain affecting older individuals or the same strain manifesting in an unusual manner due to the age of the patient.

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