Molecular and phenotypic analysis of human prion strains

https://www.neurodegenerationresearch.eu/survey/molecular-and-phenotypic-analysis-of-human-prion-strains-2/

Principal Investigators

Dr J Wadsworth

Institution

MRC Prion Unit

Contact information of lead PI
Country

United Kingdom

Title of project or programme

Molecular and phenotypic analysis of human prion strains

Source of funding information

MRC

Total sum awarded (Euro)

€ 5,640,471

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
Prion diseases are fatal neurodegenerative disorders that include scrapie in sheep, bovine spongiform encephalopathy (BSE) in cattle, Creutzfeldt-Jakob disease (CJD), Gerstmann-Straussler-Scheinker disease (GSS), fatal familial insomnia (FFI), kuru and most recently variant CJD (vCJD) in humans. Their central feature is the conversion of a normal host protein, the cellular prion protein (PrPC), to an abnormal isoform, designated PrPSc. This transition

appears to involve only conformational change rather than covalent modification and confers PrPSc with resistance to proteolYestic degradation and detergent insolubility. The marked clinical heterogeneity observed in human prion diseases has yet to be explained. However, it has been clear for many years that distinct isolates, or strains, of prions can be propagated in the same host and these are biologically recognised by distinctive clinical and pathological features. It is therefore likely that a proportion of clinicopathological heterogeneity seen human prion diseases relate to the propagation of distinct human prion strains. How this strain diversity is encoded by an apparently protein-only agent remains one of the most interesting and challenging questions in Biology. Although distinct mammalian prion strains are associated with different prion protein conformations and glycoform assembly states their precise composition and the molecular determinants of strain remain unknown. Our integrated series of research projects aim to define the molecular composition and ultra-structure of infectious prions and elucidate the molecular basis of prion strain diversity. Knowledge generated by this research is expected to have direct translational benefit by facilitating improved methods of diagnosis and therapeutic treatments for human prion disease.

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