CJD Research and Resource Centre

https://neurodegenerationresearch.eu/survey/cjd-research-and-resource-centre/

Principal Investigators

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Institution

Medicines and Healthcare products Regulatory Agency

Contact information of lead PI
Country

United Kingdom

Title of project or programme

CJD Research and Resource Centre

Source of funding information

NIHR (PRP Ad Hoc Grants)

Total sum awarded (Euro)

€ 1,473,722

Start date of award

01/04/2012

Total duration of award in years

5.0

The project/programme is most relevant to:

Prion disease

Keywords
Research Abstract

Background:
The number of individuals infected with the agent of vCJD is not known. The associated risks, which include transmission by blood transfusion and infection by blood derived proteins such as factor VIII are therefore hard to quantitate. The disease has a long silent incubation period , and a test that detected infection early would allow better informed policy decisions to be made, patients at risk to be identified definitively as infected or not and transmission by blood and blood products to be prevented. However there are very few suitable samples from patients with

vCJD. Consequently the Oversight Committee of the CJD Resource Centre at NIBSC has devised a process by which methods can be evaluated to see if they are sufficiently promising in terms of sensitivity and specificity to be given access to the few samples that exist. All likely tests at present are based on blood.

Aims:
The aim is to establish whether candidate test methods are able to identify blinded plasma or other blood samples from infected individuals with acceptable specificity, sensitivity and reliability.

Research plan:
The Resource Centre will continue to keep itself informed on developments relevant to test development through informal discusion, representatation on groups such as those of the UKBTS and ACDP, scientific meetings and, publication of assessments and updates on the web site.Collaborative research opportunities wil be pursued where appropriate. In particular possible developers will be actively followed up and methods established in house the better to understand their properties and robustness. The Resource Centre will also continue to source relevant samples to assess potential methods for specificity and sensitivity in blinded trials. This will include tissues and blood in future but the current process is to progress from panels of plasma spiked with tissues of known infectivity such as brain or spleen from patients, to blood samples from infected animals using mice, sheep cattle and primates.Normal human blood samples are used to measure specificity and finally samples from patients with clinical vCJD are provided if justified. Other samples from exposed individuals who may or may not be infected are also available including plasma from recipients of blood from donors who later went on to devlelop vCJD and haemophiliacs and other recipients of blood derived proteins. Progress is closely monitored and reviewed by the independent Oversight Committee and steps may be passed over if they consider it justified.
The Centre has materials from vCJD patients such as spleen and brain, and similar control samples from non CJD patients. It also has a collection of infected brains and spleens from sheep with clinical scrapie, cattle with BSE, experimentally infected mice and non human primates. It has acquired a number of samples from normal human donors and is pursuing overseas sources of the same type as true negative controls. It also has plasma samples from various animal models including natural scrapie in sheep, BSE in cattle, plasma from sheep infected with BSE and known to be infectious by transfusion. There is an ongoing programme to collect samples from recipients of blood from donors who later developed vCJD, from haemophiliacs notified that they are at increased risk, and recipients of immunoglobulin. The collection of samples is extensive and will continue to be supplemented including with tissues and whole blood. Two direct assay methods have completed the assessment and neither was able to identify samples from patients in a background of samples from normal donors. It is likely that methods based on amplification methods such as PMCA or QUIC will be required to reach the necessary sensitivity. These are more complex and the parameters that need to be examined to demonstrate sensitivity and speciificity largely unknown so a research effort to reproduce and establish them in house is required beginning with the expression of the recombinant protein substrate.

Research team:
The research team consists of Dr Philip MInor, Head of the Division of Virology at NIBSC, Dr Jillian Cooper and Mr Kaetan Ladhani, who have both worked on the project for many years

Potential impact:
The work is intended to identify suitable reliable tests for vCJD based on blood specimens. If such tests exist there will be a major impact on policy; if the work of the Resource Centre shows that a candidate test is not suitable this is also a vitally important finding.

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