Enhanced Creutzfeldt-Jakob Disease Surveillance in the Older Population

https://neurodegenerationresearch.eu/survey/enhanced-creutzfeldt-jakob-disease-surveillance-in-the-older-population/

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Country

United Kingdom

Title of project or programme

Enhanced Creutzfeldt-Jakob Disease Surveillance in the Older Population

Source of funding information

NIHR (PRP (ST-01-02) vCJD in the Older Population)

Total sum awarded (Euro)

€ 1,819,131

Start date of award

01/04/2015

Total duration of award in years

3.0

The project/programme is most relevant to:

Prion disease

Keywords
Research Abstract
Background: To date 177 cases of variant Creutzfeldt-Jakob disease (vCJD) have been reported in the UK, although the prevalence of detectable abnormal prion protein in the general population is estimated at 1 in 2000 people. This apparent inconsistency is poorly understood, but the possibility exists that national surveillance mechanisms could be missing some cases, particularly in older age groups, perhaps because the clinical presentation and progression of

disease in these patients is atypical of vCJD or otherwise not recognised as a prion disease. A similar situation may also exist for sCJD and perhaps other forms of prion disease. To address this, this study will seek evidence of whether some dementias in the elderly are due to CJD.

Aims: This study seeks determine whether there is otherwise unrecognised CJD (including vCJD, sCJD and other prionopathies, such as VPSPr) in the older population, and to investigate their clinical and pathological features, risk factors and whether these differ from observed clinical cases. It will do this through neuropathological screening of locally banked brain donations and through the clinical-pathological investigation of patients with atypical features of dementia (aged 65+) who are accessing local psychogeriatric services. Although focussed on Edinburgh and the Lothians, the study will determine if it is possible to establish routine systems for the diagnosis and surveillance of CJD amongst people with atypical features of dementia in the older population, that could be extended to the rest of the UK.

Research plan/method: The study will involve the neuropathological screening of Edinburgh Brain Bank donations from patients in the 65+ age-group for evidence of prion disease. Specimens will undergo standard neurological disease histopathology, prion protein immunocytochemistry, biochemistry and genotyping for evidence of prion disease. The second approach will involve local neurology and psychogeriatric services. Patients 65+ presenting with atypical features of dementia will be offered clinical examination and review, including MRI investigation, and prion protein genotyping, with an epidemiological risk factor questionnaire also undertaken. Further follow up will be undertaken for all patients to assess progress and post-mortem investigations in the event a patient dies. Research findings will be disseminated at regular intervals through meetings and peer-reviewed publication.

Research team: The NCJDRSU is an internationally recognised centre for the diagnosis of human prion disease. The Edinburgh Brain Bank is part of the UK Brain Bank Network, providing high quality post-mortem materials for diagnosis and research into disorders of the brain and nervous system. The research team brings together leading members of staff from both institutions, with substantial expertise in prion disease surveillance and clinical and laboratory research in neurology, neuroradiology, neuropathology and prion protein biochemistry in relation to dementing illness. Researchers also contribute to local clinic-based neurology services for patients as part of the University of Edinburgh Division of Clinical Neurosciences. Together with NHS psychogeriatric services the staff diagnose, treat and manage patients with dementia, across southern Scotland.

Potential impact: This study will identify CJD in the study population, and pilot a method by which enhanced methods of case ascertainment can be integrated into routine diagnostic and surveillance practice. In doing so the study will raise local awareness of CJD in older patients, with implications for the diagnosis of patients with atypical features of dementia, their treatment and care, and, as the older population has a relatively high frequency of medical interventions

compared to the young, for informing CJD-specific public health interventions in this age group. The study will also help generate hypotheses on the relationship between prion and other neuro-degenerative diseases that could be tested in other research cohorts and service settings.

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