Thank you to everyone who has taken part, and for your continued involvement with the study. Without your help and support this study would not be possible.

The Prion Surveillance Study

Welcome

Season’s greetings, and a very warm welcome to the Prion Surveillance Study 2019 newsletter. Thank you very much for your continued involvement with the study.

Changes to the Study Team

At the start of 2019 we welcomed a new senior epidemiologist to the Edinburgh team, Dr Lovney Kanguru. Lovney has a clinical and public health background and joins the team from the Usher Institute at the University of Edinburgh. Lovney is managing the study and is working closely with our research nurse, Kudzai.

The Prion Surveillance Study

Variant Creutzfeldt-Jakob Disease (vCJD) is a very rare disease which is caused by an abnormality in a naturally occurring protein called the prion protein, the presence of which can be detected in certain body tissues. A total of 178 cases of vCJD have ever been reported in the UK; the last UK patient was reported in 2016.

The prion surveillance study aims to find out whether there is evidence of the abnormal prion protein that is associated with vCJD in the blood and body tissues of certain primary immunodeficiency patients who were exposed to UK sourced immunoglobulin between 1996 and 2000.

Participation in the study involves, with consent:

- Donating a blood sample once every two years for storage for future testing when such a test becomes available.
- An annual telephone call from the research nurse in between.
- Donation of available tissue from previous surgery (for example, the tissue left over from biopsies) or from post-mortem examination, for testing for abnormal prion protein.

The study has been running for over 12 years since its start in 2006.

Study Progress in 2019

Over the past year Kudzai has been meeting and speaking with participants as part of the biennial review and telephone follow-up. During the follow-up, we provide patients with any new study related information and an opportunity to ask questions. In addition, we check for any recent surgery/operations, blood/blood products received and any changes to the ongoing immunoglobulin treatment, as part of the ongoing review of blood transfusions and surgery.
We have also been busy with recruitment of new immunology centres to join the study. We are pleased to announce that we have recruited one new centre and a further 5 centres have expressed an interest.

Since the study started in 2006 to date we have recruited 79 participants, with 13 centres currently participating. In total, study participants have donated:

- 418 blood samples over this time. The blood samples are currently stored with NIBSC for future testing, when such a test becomes available.
- 231 tissue samples. These include from 5 participants who died having generously donated their tissues for this research.

To date, there has been no evidence of prion infection in the tissues tested and no patients have shown symptoms suggestive of prion disease.

**Conference Presentations and Published Report**

Also this year, the study received a warm response at two conferences that we participated in: the Immunology & Allergy Nurses (IANG) Conference that was held in Leicester in May and the UK Primary Immunodeficiency Network (UKPIN) Biennial Conference that took place in Liverpool earlier this December.

Our latest Annual Prion Surveillance Study steering group report has been published and is available along with other information about the study on the National CJD Research & Surveillance Unit website at: https://www.cjd.ed.ac.uk/projects/prion-surveillance-primary-immunodeficiency-patients. The same report is also available on the patient group PID UK website and can be accessed at: http://www.piduk.org/whatarepids/treatment/immunoglobulinreplacmenttherapy/prionsurveilancestudyreport2019

**Over the next year…**

We hope to recruit more centres into the study, and Kudzai will continue to meet with new as well as existing study participants. We look forward to talking about the study with you and in the meantime we wish you a Happy Christmas and New Year 2020!

With best wishes,
Anna, Lovney and Kudzai

The NCJDRSU, Edinburgh

**Contact details**

If you would like to talk with us about the study please contact your local immunology team or the NCJDRSU.

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