



Newsletter No. 8

December 2023

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 2023 newsletter. Thank you for your continued involvement with the study.

The Prion Surveillance Study

The Prion surveillance study has now been running for 17 years since it commenced in 2006. It **aims** to find out whether there is evidence of the abnormal prion protein that is associated with variant Creutzfeldt Jakob Disease (vCJD) in the blood and body tissues of primary immunodeficiency patients exposed to UK sourced immunoglobulin between 1996 and 2000.

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.

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.

Summary

To date, 80 participants have taken part in the study, with 12 centres currently participating. In total, the study participants have donated:

- 464 blood samples over this time, currently stored with National Institute for Biological Standards & Control (NIBSC) for future testing, when such a test becomes available.

Contact details

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

Address:

National CJD Research & Surveillance Unit, Western General Hospital, Edinburgh



The NCJDRSU, Edinburgh

You can telephone Kudzai:
0131 537 2128 or
07557169011

Or email us:

kudzai.karekwaivanane@nhs.scot

- 260 tissue samples. These include from 8 participants who have died and generously donated their tissues for our research

There has been no evidence of prion infection in the tissues tested and no patients have shown symptoms of prion disease to date.

Over this past year

Our research nurse has been busy with the patient face-to-face visits and telephone follow ups. The research team has also participated in several conferences, where the study was met with a warm response:

- The Immunology & Allergy Nurses' conference (IANG), 15th -16th May 2023, Cardiff. [Oral presentation]
- International Society of Blood Transfusion (ISBT) congress, 17-21st June 2023, Gothenburg, Sweden. [Poster]
- British Blood Transfusion Society (BBTS) conference, 10th -12th October 2023, Harrogate, UK [Oral presentation & Poster]
- Prion 2023 conference, 16th -19th October 2023, Faro, Portugal [Oral presentation & Poster]
- British Society for Immunology- Clinical Immunology Professional Network (BSI-CIPN) Conference, 4th-5th December 2023, Belfast [Oral presentation]

Published Report

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>.

A report of the study was also published on the Immunodeficiency UK, website in August this year.

Over the next year....

Our research nurse, Kudzai, will continue with the face-to-face visits, telephone follow ups, and the recruitment of new eligible immunology centres and new patients. In the meantime, we wish you a Happy Christmas and New Year 2024!

With best wishes,
Richard, Lovney and Kudzai

