



Newsletter No1
May 2016

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.

In 2015 this study was transferred from Manchester to Edinburgh. A year later, we would like to update you on progress and provide you with information that we hope you find interesting.



The Prion Surveillance Study

THIS STUDY STARTED in 2006, and aimed to find out whether any evidence of prion infection could be found in antibody deficient patients who received certain UK-sourced immunoglobulin products between 1996 & 2000. The products were made from plasma from UK donors and have been infected with a particular type of prion that causes variant Creutzfeldt-Jakob Disease (vCJD).

VARIANT CJD is a very rare neuro-degenerative disease. Sometimes infection can be detected in certain body tissues, even before the person becomes ill. We think that most cases of vCJD have been caused by eating bovine spongiform encephalopathy (BSE) contaminated meat. However we now know that infection may be spread through blood transfusion or treatment with certain plasma products.

PATIENTS INVOLVED IN THE STUDY are followed up over several years, allowing for any available tissue (for example, the tissue left over from routine biopsies) to be tested for the abnormal prion protein that causes prion disease, and blood samples to be taken for when a blood test becomes available. Participants in the study can also agree to donate tissues obtained from a post-mortem, as this is the best way to collect samples that help us confidently state whether prion infection has taken place.

FOLLOWING THE RETIREMENT of Dr Matthew Helbert in 2015, management of the study transferred from Manchester to Edinburgh, and is now led by Dr Anna Molesworth (PhD) working with the team at the National Creutzfeldt-Jakob Disease Research & Surveillance Centre (NCJDRSU) in Edinburgh.

THE STUDY CONTINUES TO AIM TO identify whether there is evidence of abnormal prion protein/vCJD in the blood and/or body tissues of primary immunodeficiency patients exposed to UK sourced immunoglobulin between 1996 and 2000.



Chief Investigator,
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Contact details:

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

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Summary

- Approximately 175 immunodeficiency patients are thought to have been exposed to UK sourced immunoglobulin between 1996 and 2000.
- Map of current recruitment sites.



- So far, 77 patients registered in 16 immunology centres have been involved in the study.
- All participants had received UK sourced immunoglobulin, with 9 known to have been treated with implicated batches.
- To date, no evidence has been found to show prion disease. This means that no patients have shown symptoms of prion disease, nor is there any evidence of prion infection in the tissues tested.
- These results are reassuring but it is very important that we continue to monitor patients over the long term as we know that prion disease may take many decades to develop after exposure.
- There is no blood test that can reliably tell us if someone is infected before they develop symptoms of disease. All study participants have agreed to donate blood for storage for future testing when such a test becomes available.
- At the time of writing a total of 178 cases of vCJD have been reported in the UK, including three transfusion-associated cases. A further 52 cases have been reported in other countries.

Over the next year....

Emma Hughes, our newly appointed study research nurse who is based in Edinburgh, has taken over from Cathy Bangs and will be visiting patients who are already involved in the study, as well as inviting new centres and participants who are eligible to take part in the study to join. We look forward to meeting with you!