



Patient Information Sheet:

National Prion Monitoring Cohort

(Note: this information sheet is for the first strata – symptomatic patients with confirmed disease)

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A longitudinal observational study of all patients diagnosed with or at high risk of developing human prion disease

You are being invited to take part in a research study. Before you decide it is important for you to understand why the research is being done and what it will involve. This information sheet explains the whole study. It may help you decide first whether you wish to take part at all, and, if you do want to take part, how much follow-up you would be comfortable with. This will depend on your feelings about the different types of follow-up assessments. Please take time to read the following information carefully, and discuss it with others if you wish. Ask us if there is anything that is not clear, or if you would like more information. Take time to decide whether or not you wish to take part.

What is the purpose of the study?

The purpose of this study is to collect information about prion disease. No treatment will be given specifically as part of this study but you would be free to take whatever treatment you and your doctor felt appropriate. By closely documenting how prion disease develops, we hope to be in a better position to determine the effects of any future drugs and understand more about prion disease.

Why have I been chosen?

You are being invited to take part in this study because you have a prion disease. You will have already had some investigations for this. People who are likely to have inherited prion disease may have already had a gene test or may have family members who have had a gene test. Those who are likely to have variant CJD may have had a tonsil biopsy test. Those who are likely to have sporadic CJD may have received a firm diagnosis on clinical grounds together with a number of tests such as a brain biopsy.

Do I have to take part?

It is entirely up to you to decide whether to take part or not. If you do decide to take part you will be given this information sheet to keep and be asked to sign a consent form. Even after you have decided to take part, you are free to withdraw at any time without giving a reason. A decision to withdraw, or a decision not to take part, will not affect the standard of care you receive.

What will happen to me if I take part?

You will have clinical assessments at least every 6 months and at most every 6 weeks – it would be up to you how often you would be willing to be seen. If you live a long way from London and would prefer to be seen locally, we will arrange for the study staff from the National Prion Clinic to visit you at home for follow-up visits. Otherwise, you will have 3 weekly – 6 monthly clinical assessments at the National Prion Clinic in London. If you decide to have the clinical assessments in London, the travel costs for you and up to two relatives (including the cost of transport, overnight stay in a shared room at a hotel close to the National Prion Clinic and necessary food) will be paid for by the study.

We will make telephone contact at 1-2 weeks following our first visit to enquire about clinical changes, thereafter we will contact at 1-2 weekly intervals until the first review visit. After the first review the frequency of telephone contact will be altered according to changes in symptoms. If there are only minor changes in symptoms at this point the use of telephone contact may be reduced or stopped completely.

What tests and examinations are required?

- *Neurological examination*
A doctor will carry out a standard neurological examination to see how you are doing clinically. This will be repeated at each follow-up visit to help judge how the disease develops.
- *Level 1: Standard neurological assessments*

There are a number of more detailed neurological assessments that we would also like to carry out as part of this study. Many of the tests will be familiar to you and involve asking you to carry out some tasks based on words, letter or numbers. **We appreciate not everybody will want to have all the different assessments and you will be free to refuse any test you do not want.** A study doctor will explain the assessments to you.

Level 2: Optimal assessments

- Visual recording of neurological examination

We would like to visually record the neurological examination. This will enable your progress to be reviewed by doctors who are not involved with your care so that they can assess objectively how the disease develops. A unique study number and date of birth, but not your name, would be used to identify the visual recordings that would be kept securely like your hospital notes. Due to some of the tests that are part of the neurological examination, such as eye tests, we are not able to hide your face in the visual recording so it will be visible. **We appreciate not everybody will want to have visual recordings taken during the neurological examination and you will be free to refuse the visual recording.** A study doctor will explain the neurological examination to you.

- MRI scan, EEG, EMG and NC

We would like to do some special tests that will help to better understand prion disease. These include an MRI brain scan, an electroencephalogram (EEG), an electromyography (EMG) and nerve conduction (NC). **We appreciate not everybody will want to have all the different tests and you will be free to refuse any test you do not want.** Separate information sheets are available about the MRI scan, EEG, EMG and NC.

- Blood samples

A 50ml blood sample, equivalent to 8 teaspoons, will be taken – some will be used immediately for blood tests and some will be stored. New tests to look for progression and diagnosis of prion disease are being developed and storing these samples will enable these tests to be done later during the study or after the study to help research into prion disease. We are therefore asking for you to agree that blood samples can be stored and used in future for tests relevant to human prion disease. No DNA testing will be done with the blood sample. You will not benefit financially from these new tests – for example, if they were developed commercially - although you would be able to have the results after the study is completed. If you decided to withdraw from the study, and wanted these stored samples to be destroyed, we would respect your views. **We appreciate not everybody will want to have a blood sample taken and you will be free to refuse the blood test.**

We will discuss with you at each visit which of these assessments you would feel comfortable with and you would be able to manage. You may be asked by the clinical team to keep a daily diary recording symptoms and the doctor will discuss this further with you.

What are the possible disadvantages and risks of taking part?

As no drugs are prescribed as part of this study, the only potential risks are those associated with investigations, which are minimal. You will have a number of additional tests and hospital visits in addition to those necessary for normal care.

What are the potential benefits of taking part?

The information we get from this study may help us to treat future patients with prion disease better.

Is there anything else that I may be asked to do?

In the event of your death, additional information about the prion protein in the brain may help explain results from this study. This would require a post-mortem. Consent to a post-mortem examination in the event of your death is **not** required for participation in this study. However, your doctors may wish to discuss this with you. This can be discussed either now, or at a later time, or not at all if you feel it is inappropriate. Please tell the nurse or doctor what you would prefer.

You may be considering giving, or you may have already given, blood for DNA tests to other studies of human prion disease. Rather than repeating these tests we are asking if you would agree to any genetic information from these tests (such as mutations in different genes) being linked to this study, in order to investigate and better understand how different people with prion disease might respond to treatment. This linking would only be done anonymously through your study number, so you would not get any results from these extra analyses.

Will my taking part in this study be kept confidential?

The study will be conducted in compliance with the Data Protection Act, 1998. All information that is collected about you during the course of the study will be kept strictly confidential but it will not be completely anonymous. All information about you, including samples will be stored only by a study number, your date of birth and initials, and not by your name. Names will not be used in any of the study records or samples. When you consent to take part in this study you also agree to allow authorised staff from the MRC Prion and Clinical Trials Units to inspect your medical records to monitor the study. In no circumstances will your name or address be disclosed outside the clinic. We would like you to let us tell your GP and any other doctor looking after you that you have entered this study. Also, we will seek your agreement to be able to telephone you or your family if you miss clinic visits or we are concerned about your health. However, if you do not want us to telephone you or your family, this will be respected and you can still join the study. Any blood and other samples taken from you in the course of this study may be used for further research in this area. By signing the consent form you are authorising the use of these tissues or body fluids for these future tests.

What will happen to the results of the study?

We will regularly review data from the study and publish results in peer reviewed journals. A summary of any published results will be provided for you if you wish. You will not be personally identified in any report or publication on this study.

Who is organising and funding the research?

The Medical Research Council (MRC) Prion and Clinical Trials Units are organising the National Prion Monitoring Cohort. The Department of Health is funding the study. The doctors looking after you will not receive any additional payments if you enter the study. Your hospital will be reimbursed for any extra tests done as part of the study.

Who has reviewed the study?

The study has been reviewed by the Scotland A Multi-centre Research Ethics Committee.

Contact for further information

Please address any further questions to Simon Mead, Consultant Neurologist and Clinical Co-ordinator; or Professor John Collinge, Clinical Director, at the National Prion Clinic:

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Thank you for taking the time to read this information sheet