



Relative/carer/consultee Information Sheet:

National Prion Monitoring Cohort

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

Your relative/ward is being invited to take part in a research study. Before you decide whether you agree to their participation in this study it is important for you to understand why the research is being done and what it will involve for your relative/ward. This information sheet explains the whole study. It may help you decide first whether you wish for your relative/ward to take part at all, and, if you do want them to take part, how much follow-up you would be comfortable with them receiving. 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 your relative/ward 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 as part of this study but your relative/ward would be free to take whatever treatment they and their doctor felt appropriate. By closely documenting how the 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 has my relative/ward been chosen?

Your relative/ward is being invited to take part in this study because they have (or are likely to have) a prion disease. Your relative/ward 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.

Does my relative/ward have to take part?

It is entirely up to you to decide whether you would like your relative/ward to take part or not, however, you should only agree for your relative/ward to participate in this study if you think they would not have refused to take part. If you do decide that you would like them to take part you will be given this information sheet to keep and be asked to sign an agreement form (or a consent form if your relative/ward is aged under 16 years). Even after you have decided to allow your relative/ward to take part, you are free to withdraw your agreement/consent 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 your relative/ward receives.

What will happen to my relative/ward if they take part?

Your relative/ward will have clinical assessments at least every 6 months and at most every 6 weeks – it would be up to you and them how often the assessments would be. If your relative/ward lives a long way from London and you would prefer them to be seen locally, we will arrange for the study staff from the National Prion Clinic to visit your relative/ward at home for follow-up visits. Otherwise, your relative/ward will have 6 weekly – 6 monthly clinical assessments at the National Prion Clinic in London. If you decide for your relative/ward to have the follow-up visits in London, their travel costs

and those for up to two relatives/carers (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. We may wish to interview carers for around 30-60 minutes to find out about how the patient's illness impacts on home life. The interview will be conducted by a clinical nurse specialist. The interview will be audio recorded for transcription purposes, then destroyed. Information taken from the interview will be held confidentially and may be used anonymously in a scientific publication.

What tests and examinations are required?

Neurological examination

Your relative/ward will undergo a clinical neurological examination to see how they are doing clinically. This examination 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 your relative/ward and involve asking them to carry out some tasks based on words, letters or numbers. **We appreciate not everybody will want their relative/ward to have all the different assessments and you will be free to refuse any test you do not want them to have.** 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 relative/ward's progress to be reviewed by doctors who are not involved with their care so that they can assess objectively how the disease develops. A unique study number and date of birth, but not your relative/ward's name, would be used to identify the visual recordings that would be kept securely like their 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 relative's face in the visual recording so it will be visible. **We appreciate not everybody will want their relative/ward to have visual recordings taken during their 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 from your relative/ward – 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 from your relative/ward can be stored and used in future for tests relevant to human prion disease. No DNA testing will be done with the blood sample. Your relative/ward will not benefit financially from these new tests – for example, if they were developed commercially - although you and your relative/ward would be able to have the results after the study is completed. If you decided to withdraw your relative/ward from the study, and wanted their stored samples to be destroyed, we would respect your views. **We appreciate not everybody will want their relative/ward to have a blood sample taken and you will be free to refuse them having a blood test.**

We will discuss with you at each visit which of these assessments you feel would be appropriate for your relative/ward and they 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 my relative/ward taking part?

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

What are the potential benefits of my relative/ward 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 my relative/ward may be asked to do?

In the event of your relative/ward's 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 relative/ward's death is **not** required for participation in this study. However, your relative/ward's 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.

Your relative/ward may be considering giving, or they 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 your relative/ward's 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 relative/ward's study number, so they would not get any results from these extra analyses.

Will my relative/ward's 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 your relative/ward during the course of the study will be kept strictly confidential but it will not be completely anonymous. All information about your relative/ward, including samples will be stored only by a study number, date of birth and initials, and not by name. Names will not be used in any of the study records or samples. When you consent to your relative/ward to take part in this study you also agree to allow authorised staff from the MRC Prion and Clinical Trials Units to inspect your relative/ward's medical records to monitor the study. In no circumstances will your relative/ward's name or address be disclosed outside the clinic. We would like you to let us tell your relative/ward's GP and any other doctor looking after them that they have entered this study. Also, we will seek your agreement to be able to telephone you or another of your relative/ward's relatives/carers if they miss clinic visits or we are concerned about their health. However, if you do not want us to telephone you or another relative/carer, this will be respected and your relative/ward can still join the study. Any blood and other samples taken from your relative/ward 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 your relative/ward's 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 and your relative/ward if you wish. Your relative/ward will not be identified in any report or publication on this study.

Who is organising and funding the research?

The 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 your relative/ward will not receive any additional payments if your relative/ward enters the study. Your relative/ward's 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 Committees.

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