Request for Creutzfeldt-Jakob Disease Surveillance (Field Survey)

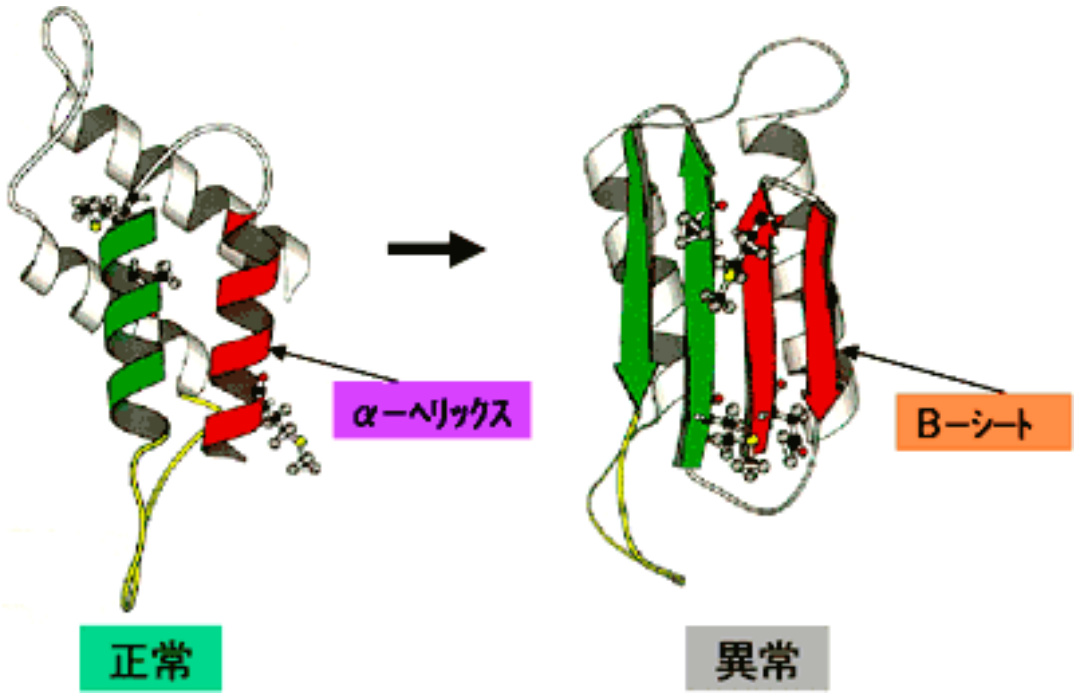
Grant-in-Aid for Health, Labour and Welfare, Research Project for Overcoming Intractable Diseases
Research on Surveillance and Prevention of Prion Diseases
" Team Leader: Dr. Hidehiro Mizusawa,
Chairman of the Creutzfeldt-Jakob Disease Surveillance Committee
Hidehiro Mizusawa,
President, National Center of Neurology and Psychiatry

1. Introduction

This document is intended to assist those who are considering participating in this study in understanding the content of the study as explained to them by the principal investigator or subinvestigator.
Please decide whether or not to participate in the research after receiving an explanation of the research and understanding the contents of this document. We ask that your decision to participate in the research be made of your own free will. If you decide not to participate in the research, we guarantee that you will not be disadvantaged in any way. Also, please understand that the results of this research may result in intellectual property rights, such as patent rights, in the future, but that these rights do not belong to you, the research participant.
If you have any questions or concerns about the research, please do not hesitate to ask us. ◦

★Prions and Prion Disease★

In some diseases, one of the many proteins that exist in the brain and other parts of a healthy person's body changes its shape and properties for some reason, accumulates in the brain, and causes loss of brain function.
In the past, it was believed that the brain and other parts of the body were infected by some pathogen, but after long research, it was discovered that the disease-causing agent (infectious agent) is a protein. The disease-causing agent is called a "prion. The disease in which these "prions" accumulate in the brain is called "prion disease.
What role and function the normal prion protein has in the human or animal body before it changes form is still not well understood.
However, it is known that normal prion protein has a helical structure as shown in the left side of the figure below. The helical structure of the normal prion protein, as shown on the left side of the figure below, is known to have a helical structure.
It is known that the prion protein somehow changes into the sheet structure shown in the right side of the figure below and accumulates in the brain and other parts of the body as an abnormal prion protein that is very difficult to degrade.



It is known that an abnormal prion protein that has changed its shape to a sheet form has the ability to change neighboring proteins to the same form. This is called "propagation. Through this propagation, the increased number of abnormal prion proteins accumulates in the brain and other parts of the body.

Prion diseases are also found in animals such as cattle and sheep. Known human prion diseases include Creutzfeldt-Jakob disease, Gerstmann-Streisler-Scheinker disease, and Kreutzfeldt-Jakob disease, in which genetic abnormalities have been reported.

★Research on prion diseases★

It has been nearly 30 years since the cause of prion disease was first identified as an abnormal prion protein.
At present, there are no proven effective treatments for prion diseases, but medical science is advancing day by day and research is progressing.

In the future, when developing drugs and treatments, it will be necessary to determine what the natural progression of the disease is in the first place in order to determine if the drugs and treatments are improving the disease or slowing the progression of the disease. In other words, in order to develop a treatment or prevention for the disease, we first need to know the natural time course of the condition of the patient who has the disease. This kind of research is called natural history research.
By conducting this national survey study of the natural history of prion diseases, it will be possible to evaluate whether or not a potential medication or treatment, when found, will actually help treat the disease.

Prion diseases are extremely rare, affecting only about 250 people per year. We would like to ask for the cooperation of all patients who are known to have prion diseases, and in order to gather as much information as possible about the disease, we would like to investigate the progress of the disease through medical examinations and telephone interviews.

2 ・Title of this study

Title of Study: "Survey Study on Prion Disease Surveillance and Prevention of Transmission"
National Survey Study on Prion Disease Surveillance and Natural History
-Study 1 (Creutzfeldt-Jakob Disease Surveillance Study (Field Survey))
Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History)) - This study is being conducted with the approval of the Ethics Committee of the Center and the President.

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3 ・**Purpose and Significance of this Study**

Creutzfeldt-Jakob disease (hereinafter referred to as CJD) and related diseases are progressive, intractable diseases of the nervous system, and in recent years, a new type of CJD (called variant CJD), which may be transmitted from bovine spongiform encephalopathy (BSE), has emerged as a major social problem. However, the mechanisms of pathogenesis, preventive measures, and treatme nt of these diseases are still unknown. The purpose of this surveillance survey is to clarify the trends of these diseases and to help prevent infection, develop new diagnostic methods, and develop effective treatments.

The "Prion Disease Surveillance and Infection Prevention Research Group" launched the CJD Surveillance Committee in FY1999 and has been conducting surveillance of patients with CJD based on the clinical survey forms submitted to each prefecture by patients under the Research Project to Combat Intractable Diseases funded by the Ministry of Health, Labor and Welfare of Japan, with the consent of the patients. We have been conducting surveillance surveys with the consent of patients suffering from CJD based on the individual clinical survey forms submitted by patients to each prefecture. Currently, the entire country is divided into 10 regional blocks, and each block has its own surveillance committee member, who, with the cooperation of CJD specialists assigned to each prefecture, directly examines patients' medical conditions.

—Study 1 (Creutzfeldt-Jakob disease surveillance study (field study))—

Creutzfeldt-Jakob disease (CJD) and related diseases are progressive, intractable diseases of the nervous system, and have become a major social problem in recent years with the outbreak of a new type of CJD (called mutant CJD), possibly transmitted from cattle affected by bovine spongiform encephalopathy (BSE). Prion diseases are very rare, and there is currently no proven effective treatment. The mechanism of pathogenesis of these diseases is unknown, and preventive measures and treatments have not yet been developed.
Therefore, we are conducting this surveillance survey in order to clarify the trends of disease outbreaks and to help prevent infection, develop new diagnostic methods, and effective treatments.

—Study 2 (Creutzfeldt-Jakob disease surveillance study (natural history study))—

Prion diseases are very rare and currently have no proven effective treatments.
In order to develop a treatment for this disease in the future, it is necessary to gather as much information as possible about the disease in order to know how the disease progresses in the first place and to compare the efficacy of candidate drugs and other treatments.

Therefore, we would like to conduct this study to determine how the physical functions of patients

with this disease change over time, and we would like to hear from family members who are caring for patients.

4 · Method of conducting this research and period of participation

—Study 1 (Creutzfeldt-Jakob disease surveillance study (field study))—

Patients who have been diagnosed as possible or probable according to the diagnostic criteria of the WHO and the Ministry of Health, Labour and Welfare's "Research Group on Surveillance and Prevention of Prion Diseases" and who have agreed to participate in this study will be included. Any gender and any age are acceptable.

Based on the CJD Surveillance Committee established in 1999, the entire country will be divided into 10 regional blocks, each with its own surveillance committee member, and furthermore, patients' medical conditions will be investigated directly under the supervision of a CJD specialist assigned to each prefecture.

Specifically, we will examine the patients and examine their test results at the same time, and ask that the records be used for the purpose of the survey and research.

The survey will be conducted based on the individual clinical survey form submitted by the patient to the prefectural government and the test request form submitted to the laboratory (Nagasaki University, Tohoku University, etc.) when requesting spinal fluid and genetic tests, and with the consent of the patient suffering from CJD (and his/her proxy).

In requesting this, we promise the following

1) Criteria for participation in the study

This study invites participants who meet all of the following criteria to participate.

●Primary selection criteria

1) Those who have been suspected of having prion disease by their primary physician.

2) Those whose primary physician has not ruled out the possibility of prion disease, although other diseases are likely to be present.

In addition to the above, your physician will determine whether you are eligible to participate in the study based on the results of your medical examination and tests. In some cases, you may not be able to participate in the research even after you have given your consent. Also, please note that even during the course of your participation in the study, if your physician determines that it would be difficult for you to participate in the study, your participation in the study may be terminated.

2) Drugs/medical devices/therapeutics to be used in the research

This research is an observational study with the main objective of diagnosing whether or not a patient has prion disease, and therefore does not include any treatment or therapeutic drugs.

3) Research period and schedule

No hospitalization or outpatient visits are required to participate in this study.

Your attending physician will simply examine you and fill out a survey form with test results and examination findings related to your disease.

4) The tests to be conducted

This research is a study to collect medical information obtained through normal diagnosis, treatment, and examinations. We will only ask you to provide us with the results of tests performed by your doctor to diagnose your disease.
The test results will be entered on a survey form without your name and with only your age, date of birth, prefecture, and medical record number, and will be sent to the Surveillance Office and kept in strict confidence.

5) Information on available genetic counseling

Genetic counseling is available for this study. If you or your family members have been diagnosed with hereditary (familial) prion diseases, please contact the study office if you or your family members wish to receive such counseling. Our expert genetic counselors will be happy to assist you.

—Study 2 (Creutzfeldt-Jakob disease surveillance study (natural history study))—

1) Criteria for participation in the study

①Patients who have been diagnosed as "possible" or "probable" according to the diagnostic criteria of the WHO and the Ministry of Health, Labour and Welfare's "Prion Disease Surveillance and Prevention Research Group" and who have agreed to participate in this study will be included.
②Patients aged 20 years or older, regardless of gender, will be included in the study.

2) Drugs/medical devices/therapies to be used in the study

This research is an observational study with the main objective of diagnosing whether or not a patient has prion disease, and therefore does not include any treatment methods or therapeutic drugs.

3) Research period and schedule

Participation in this study does not require hospitalization or outpatient visits.
First, your attending physician will examine you and fill out a survey form with test results and examination findings related to your disease.
Patients who are eligible for this research study will be asked to undergo a monthly medical examination by a physician or a telephone survey by an investigator (telephone survey).
(The telephone survey will be answered by a caregiver or a proxy if the patient is unable to respond to the survey).
Other than that, we will not ask you to do anything else specifically for this study. We will refer to the results of tests performed during a normal medical examination.

4) Tests to be performed

This research is a study in which normal diagnosis, treatment, and tests will be performed, and the medical information obtained in the course of these tests will be collected over time. Your doctor will simply observe and inform us of any changes in your disease condition.
The observations will be entered on a survey form without your name and with only your age, date of birth, and prefecture, and will be kept strictly confidential within the National Center of Neurology and Psychiatry, separated from your personal information. (Personal information such as the patient's name and address are not included in the data. The e-mail address and telephone number of the patient or surrogate will be used in a completely separate table, so the investigators will not know the date of birth or address either).

If we wish to use the data you provide for other research purposes, we will apply for a new ethics review and use the data only after approval.

5) Information on available genetic counseling

Genetic counseling is available for this research. If you have been diagnosed with hereditary (familial) prion disease and you or your family members wish to receive such counseling, please contact the study office. Our professional genetic counselors will be happy to assist you.

5. burdens, risks, and benefits that may arise from your participation in the study

Study 1 (Creutzfeldt-Jakob disease surveillance study (field study))
Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (natural history survey) -

1) Burden and risk that may occur

(1) Adverse events

All unwanted or unintended injuries or illnesses or signs thereof (including abnormal laboratory values) that occur as a result of participation in the study are referred to as "adverse events. This research is a study in which normal diagnosis, treatment and tests are performed and medical information obtained in the course of such diagnosis, treatment and tests is collected, and no adverse events will occur to you as a result of your participation in the study.

(2) Other burdens and disadvantages

This research is to collect medical information obtained through normal diagnosis, treatment, and examination, and there will be no direct disadvantages to you as a result of your participation.

2) Anticipated benefits

This research is a study to collect medical information obtained in the course of normal diagnosis, treatment, and examinations.
However, by clarifying test results and the progress of the disease, it may contribute to the development of better treatment and diagnostic methods.

3) When to discontinue the research

1) If it becomes clear after your participation in the research that you should not participate in the research
2) If the principal investigator or others decide to discontinue the entire study.

6. withdrawal of consent after participation in the study

-Study 1 (Creutzfeldt-Jakob Disease Surveillance Study (Field Survey))
Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History)
Your decision to participate or continue in this study is of your own free will. You will not be disadvantaged in any way if you decline to participate in this study. Even once you have given your consent to participate in the research and the research has begun, you may stop participating at any time.

You will not be disadvantaged in any way.

7.Disclosure of Information on Research

Study 1 (Creutzfeldt-Jakob disease surveillance study (field survey))
Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History))
The characteristics of prion diseases in Japan as elucidated by these studies are published semiannually on the website of the Prion Disease Surveillance Study. The results of the natural history study are also published on the Surveillance Research Committee.
The results of this research will also be presented at academic conferences and in academic papers.

8. to obtain or view the research protocol and methodology documents

-Study 1 (Creutzfeldt-Jakob disease surveillance study (field survey))
Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History)) -

The research protocol and materials related to the methodology of the research may be viewed, provided that this does not interfere with the protection of the personal information of other participants in the research or with the originality of the research. If you wish to do so, please contact us at the address listed at the end of this document.

9. Handling of Personal Information, etc.

-Study 1 (Creutzfeldt-Jakob Disease Surveillance Study (Field Survey)-)

Your name will not be entered on the survey form completed by your attending physician, and you will be distinguished from others by your initials, gender, date of birth, and medical record number. The data related to the research, such as your test results and medical information, that you fill out on the same survey form will likewise be anonymized and managed without knowing your name.

These anonymized data will be reviewed by the Surveillance Committee and will be used for the purpose of the Creutz Feldt disease (prion disease) study.

These anonymized data are reviewed by a surveillance committee to determine if you have Creutzfeldt's disease (prion disease).

The data is stored in a data warehouse, which is also used by other financial and medical institutions, and which is protected by a strict information security policy.

The data is stored digitally (encrypted in computer language) in a data warehouse that is also used by other financial and medical institutions.

The information will be digitized (encrypted in computer language) and stored in a place where strict information security is guaranteed (called a data warehouse, which other financial institutions and medical institutions also use privately).

In addition, your information may be accessed by persons involved in the clinical research (personnel authorized by the principal investigator), the MHLW and its related agencies, and ethics committees to ensure that this clinical research is conducted properly. However, these parties are obligated to maintain confidentiality and your personal information will not be misused.

-Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History Study)-)

Your name will not be on the survey form completed by your attending physician, but only your initials, gender, date of birth, and medical record number to distinguish you from others. We will not ask you or your surrogate directly about your medical condition, nor will we ask you or your surrogate directly about your medical condition when we subsequently interview and record your medical condition with your primary care physician over time.

There will be no identifying information on the form that you or your surrogate will fill out.

We may contact you or your surrogate by e-mail or telephone number that you or your surrogate have provided with your consent, but such e-mails and telephone numbers will be kept in a separate file from the questionnaire that describes your medical condition, and will not be stored in a separate computer system under strict control of information management.

These e-mails and phone numbers will be kept in a separate file and stored on a separate computer under strict information control, so that the association of your detailed personal information with your illness will be minimized to the greatest extent possible.

10. Methods of Storage and Disposal of Samples and Information, and Secondary Use of Samples and Information

Study 1 (Creutzfeldt-Jakob Disease Surveillance Study (Field Survey))

Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History)) -

The materials and information obtained from this study will be stored for a minimum of 5 years in the Surveillance and Natural History Research Office at the Center, using only your initials, gender, date of birth, and your hospital record number (anonymized), without using your name.

Samples and information obtained from this research will be discarded at the end of the study after confirming that your personal information is not included, but may be provided to other research institutions for secondary use with your consent.

11. conflicts of interest related to research, including funding sources and researchers

Study 1 (Creutzfeldt-Jakob Disease Surveillance Study (Field Survey))

Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History Survey)

A "conflict of interest" is a situation in which a third party may be concerned that the research is not being conducted fairly and appropriately, such as falsification of research data, favoritism toward a particular company, or continuation of research when it should be discontinued, due to financial interests with an outside party. Surveillance research is funded by public funds (Grant-in-Aid for Health, Labour and Welfare for Research on Intractable Diseases, Research on Prion Disease Surveillance and Prevention of Infection, and Government Support for Home Healthcare for Patients with Neurological Intractable Diseases, PI: Hidehiro Mizusawa, period: April 1, 2017 - March 31, 2022). No funds were provided by any specific company. The natural history research study will also be conducted fairly under the auspices of the University's principal investigator and JACOP, with scientific research grants, etc., provided by the government. This research study has been approved by the Ethics Review Committee and will be kept fair.
Conflicts of interest of the Center's researchers in this research are reviewed by the Center's Conflict of Interest Management Committee and are properly managed. The conflicts of interest of researchers at collaborating institutions have also been reviewed and confirmed to be acceptable.

12. if you have any questions regarding this study

-Study 1 (Creutzfeldt-Jakob Disease Surveillance Study (Field Survey))
-Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History))
If you or your family have any questions or concerns about this study, please do not hesitate to contact us.

If you or your family members have any questions or concerns about this study, please do not hesitate to contact the contact person at the end of this document. Please note that we may not be able to respond to your inquiry or answer your questions due to reasons such as protecting the personal information of other research participants or the intellectual property rights of the researcher.
In addition, if you have any complaints about any inconvenience caused by the implementation of this research, please contact the Complaints Office (Ethics Committee Office of the Center).

13. Financial Burden Associated with Participation in Research

Study 1 (Creutzfeldt-Jakob disease surveillance study (field survey))
Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History)
Participation in this study will not involve any additional financial burden for you compared to your regular medical care.
You will be expected to pay for your own medical examinations and medications performed by your primary care physician for diagnosis and treatment of your disease using your health insurance, just as you would if you were receiving regular medical care.

14. other treatment modalities

-Study 1 (Creutzfeldt-Jakob disease surveillance study (field study))
Study 2 (Creutzfeldt-Jakob disease surveillance study (natural history study)
This study is to collect medical information obtained in the course of routine diagnosis, treatment, and testing. Failure to participate in the study will not result in a change in treatment procedures.

15. Provision of medical care after the study is conducted

-Study 1 (Creutzfeldt-Jakob Disease Surveillance Study (Field Study)-)
This study is to diagnose people suspected of having prion disease, including you. It is not intended to provide medical care after diagnosis.
Please consult with your doctor regarding your treatment plan after diagnosis. If necessary, the Surveillance Committee of this study can provide advice to your doctor.

-Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History Study))
This study will follow the evolution of the disease in persons suspected of having prion disease, basically until they become immobile and mute, and even until death. Subsequent provision of medical care includes cooperation in autopsies. (by the pathology member of the surveillance committee).

16. handling of research results for research participants

Study 1 (Creutzfeldt-Jakob disease surveillance study (field study))
Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History)
In conducting this study, it is possible that important findings may be obtained regarding your health status.

During MRI imaging, it is possible that findings that suggest the presence of a serious disease, such as a brain tumor, may be obtained by chance. However, the MRI imaging in this study is not intended to confirm the health status of the participants and cannot be used for medical diagnosis. If you wish to have a separate diagnosis, we will refer you to a formal examination, but you will have to bear the cost of the examination, and a formal examination may again show that there is no problem. For these reasons, we would like to know in advance whether or not you wish to receive information on any findings that may arise by chance as a result of your participation in the study that may not be ignored.

17. compensation for any adverse health effects resulting from participation in the study

Study 1 (Creutzfeldt-Jakob disease surveillance study (field study))
Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History)

The main purpose of this study is for the Surveillance Committee members to gather and diagnose the information in the survey form, and this study itself will not cause any harm to your health.
In the unlikely event that any damage to your health should occur as a result of this research, there will be no special compensation in the form of medical expenses, medical benefits, or compensation. Please fully understand this point and decide whether or not to participate in the research. Please note that understanding the contents of this explanation document and agreeing to participate in this clinical research does not mean that you waive your right to claim compensation for any health damage.

18. handling of received samples and information (possible future use)

Study 1 (Creutzfeldt-Jakob disease surveillance study (field study))
Study 2 (Creutzfeldt-Jakob disease surveillance study (natural history survey)
Your test results, examination findings, and diagnostic results from the questionnaire may be provided to other institutions and used in the future to contribute to prion disease research. Of course, in this case, information that identifies you will not be included in the information provided.
The names of institutions that may be provided are the medical and university institutions to which the members of our surveillance committee belong, as well as prion disease laboratories and research organizations overseas. The research that may be conducted is related to the epidemiology of prion diseases in Japan and may be comparative research with other countries.
Such collaborative research with medical institutions would probably be publicly funded (because of the rare nature of the disease, general corporate funding is unlikely). Such research would need to be reviewed by the Center's Ethics Committee and approved by the Center before it could be conducted.
(Since it is a rare disease, funding from general companies is unlikely to be forthcoming.

19. name of the organization, name of the institution, and name of the principal investigator(s) for this study

-Study 1 (Creutzfeldt-Jakob Disease Surveillance Study (Field Survey))
Study 2 (Creutzfeldt-Jakob Disease Surveillance Study (Natural History)) - The principal investigators for both studies are
Name of the principal investigator: Hidehiro Mizusawa, President, National Center of Neurology and Psychiatry, National Institute of Neurology and Psychiatry, Japan

-Study 1 (Creutzfeldt-Jakob Disease Surveillance Study (Field Survey))
The secretariat of the Surveillance Committee was set up at the Center, and committee members from all over Japan met twice a year.
The committee will meet twice a year to diagnose prion diseases based on the information obtained. This is a stand-alone study at our center, but the committee will be composed of members belonging to the following facilities.

サーベイランス 委員会			
	氏名	所属機関 部署	役職
委員長	水澤 英洋	国立精神・神経医療研究センター	理事長
委員	山田 正仁	金沢大学医薬保健研究域医学系 脳老化・神経病態学 神経内科学）	教 授
	斉藤 延人	東京大学大学院医学系研究科 脳神経外科学	教 授
	北本 哲之	東北大学大学院医学系研究科 病態神経学分野	教 授
	中村 好一	自治医科大学 公衆衛生学	教 授
	黒岩 義之	財務省診療所	所長
	原田 雅史	徳島大学ヘルスバイオサイエンス研究部放射線科学分野	教 授
	佐藤 克也	長崎大学医歯薬学総合研究科運動障害リハビリテーション分野 神経 内科学）	教 授
	村山 繁雄	東京都健康長寿医療センター研究所 神経病理部門	部長
	太組 一朗	日本医科大学武蔵小杉病院 脳神経外科	講師・医長
	佐々木 秀直	北海道大学大学院医学研究院神経病態学分野神経内科学教室	特任教授
	青木 正志	東北大学大学院 神経内科学	教 授
	小野寺 理	新潟大学脳研究所神経内科	教 授
	三條 伸夫	東京医科歯科大学大学院 脳神経病態学 神経内科学）	プロジェクト教授
	村井 弘之	国際医療福祉大学医学部神経内科	教 授
	塚本 忠	国立精神・神経医療研究センター病院 神経内科）	医長
	田中 章景	横浜市立大学医学部神経内科	教 授
	道勇 学	愛知医科大学	教 授
	望月 秀樹	大阪大学大学院医学系研究科 神経内科学	教 授
	阿部 康二	岡山大学大学院医歯薬学総合研究科 脳神経内科学	教 授
	松下 拓也	九州大学大学院医学研究院 神経内科学	講 師

２０．Inquiries concerning this research

○ For inquiries regarding this research, please contact

4-1-1 Ogawa-higashi-machi, Kodaira-shi, Tokyo 187-8551, Japan
National Center of Neurology and Psychiatry

Phone: 042-341-2712 ext. 3131
Prion Disease Surveillance Secretariat
Tadashi Tsukamoto

Office of Surveillance Research and JACOP Natural History Research
Phone: 042-341-2712 ext. 3131 or 3133
(Available weekdays from 9:00 a.m. to 5:00 p.m.)

○ Complaint consultation

4-1-1 Ogawa-higashi-machi, Kodaira-shi, Tokyo 187-8551, JapanNational Center of Neurology and Psychiatry, National Research Institute of Neurology and Psychiatry, Ethics Committee Secretariat e-mail：ml_rinrijimu@ncnp.go.jp

【Form 1】 【Doc 6-1】

Date: 30 November 2017

Consent Document for Research Participation

Grant-in-Aid for Health, Labour and Welfare, Research Project to Conquer Intractable Diseases
Dr. Hidehiro Mizusawa, Chairperson, Prion Disease Surveillance Committee, "Investigational Research on Surveillance and Prevention of Prion Diseases

I am writing to explain the following matters concerning the "Prion Disease Surveillance (Implementation Survey)" of the "Prion Disease Surveillance and Infection Prevention Research" Group.
（I have fully explained and understood the following matters concerning the "Prion Disease Surveillance and Prevention Study" (Implementation Study) using the Explanatory Document (November 29, 2017, Version 1). I am participating in this study of my own free will

Items explained and understood
□1 Purpose and significance of this study (Explanatory document, item 3)
□2. How this research will be conducted and the period during which you will participate (Explanatory Document, item 4)
□3 Burdens, risks, and benefits that may arise from participation in the research (Explanatory Document, Item 5)
□4 Voluntary nature of participation in the study and withdrawal of consent after participation in the study (Explanatory Document, Item 6)
□4-1 That you may withdraw your consent to participate in this research at any time.
□4-2 That you will not be disadvantaged in any way in terms of treatment even if you do not participate in the research or if you withdraw your consent.
□5 Disclosure of Information on Research (Explanatory Document, Item 7)
□6. If you wish to obtain or view the research protocol and materials related to the research methods (Explanatory Document, Item 8)
□7. Handling of Personal Information (Explanatory Document, Item 9)
□8 Methods of storage and disposal of samples and information, and secondary use (Explanatory document, item 10)
□9 Conflicts of interest related to research, such as funding sources and researchers (Explanatory Document, item 11)
□10 Financial burdens associated with participation in research (Explanatory Document, item 13)
□11 Other treatment methods (Explanatory Document, item 14)
□12 Provision of medical care after the research is conducted (Explanatory Document, item 15)
□13 Handling of research results for research participants (Explanatory Document, item 16)
Do you wish to be informed when findings that cannot be ignored in terms of health are discovered by chance or when important findings are obtained?
□ Yes □ No
14 Compensation for damage to health caused by participation in research (Explanatory Document, item
15 Handling of received samples and information (possibility of future use) (Explanatory document, item

At the same time, I have fully explained and understood the National Research Study on the Natural History of Prion Disease (November 29, 2017, Version 1), which will investigate the future course of the disease, using the Explanatory Document (November 29, 2017, Version 1). I was asked to cooperate in the natural history research study by providing my clinical records, and was informed and confirmed that these clinical records would be used as basic data for the research study, that my privacy would be protected, that I could give my consent completely freely, and that I could withdraw my consent at any time even after I had given it. I confirmed these points. I understand that I will provide my e-mail address or telephone number to the secretariat for natural history research, and that I will keep the secretariat informed of any subsequent changes in my medical condition. I participate in this study of my own free will
□1 Purpose and significance of this study (Explanatory document Item 3)
□2. How the study will be conducted and the period during which I will participate (Explanatory Document, Item 4)
□3 Burdens, risks, and benefits that may arise from participation in the research (Explanatory Document, Item 5)
□4 Voluntary nature of participation in the study and withdrawal of consent after participation in the

study (Explanatory Document, Item 6)
□4-1 That you may withdraw your consent to participate in this research at any time.
□4-2 That you will not be disadvantaged in any way in terms of treatment even if you do not participate in the research or if you withdraw your consent.
□5 Disclosure of Information on Research (Explanatory Document, Item 7)

□6. If you wish to obtain or view the research protocol and materials related to the research methods (Explanatory Document, Item 8)
□7. Handling of Personal Information (Explanatory Document, Item 9)
□8 Methods of storage and disposal of samples and information, and secondary use (Explanatory document, item 10)
□9 Conflicts of interest related to research, such as funding sources and researchers (Explanatory Document, item 11)
□10 Financial burdens associated with participation in research (Explanatory Document, item 13)
□11 Other treatment methods (Explanatory Document, item 14)
□12 Provision of medical care after the research is conducted (Explanatory Document, item 15)
□13 Handling of research results for research participants (Explanatory Document, item 16)
Do you wish to be informed when findings that cannot be ignored in terms of health are discovered by chance or when important findings are obtained?
□ Yes □ No
□14 Compensation for damage to health caused by participation in research (Explanatory Document, item
□15. Handling of received samples and information (possible future use) (Explanatory document, item 18)

Based on the above understanding, please check one of the following.
I agree to both surveillance and natural history research.
I agree to the Surveillance Registry Study, but not to the Natural History Study.

【 If any of the following applies】

□ Consent by proxy

Date of Consent Date of Consent

Signature

Address

Phone Number

I have received a full explanation of the above matters concerning the "Research on Prion Disease Surveillance and Infection Prevention" in which I, the undersigned, will participate, and I understand the same. I have fully explained and understood the above matters in writing. I agree to participate in this research of my own free will.
Date of Consent Date of Consent

Signature (self-signed) (Relationship)
I have explained this research to the subject in accordance with the explanatory document for this research in order to obtain his/her consent. Date of explanation
Signature of Explanator

Institution to which the explainer belongs

Request to the attending physician
Please keep the consent document ([Form 1]) at your side to protect the patient's personal information.
To indicate that consent has been obtained, please complete the Consent Confirmation Form ([Form 2]), convert it to PDF format, and send it to the Surveillance Research Office (prion-ncnp@ncnp.go.jp).

【Form 2】 【Doc 6-2】

Prion Disease Surveillance and Prevention Group Prion Disease Surveillance (Field Surveillance)
（Agreement Confirmation Form for Prion Disease Surveillance (Field Survey and Natural History Study)

Grant-in-Aid for Scientific Research on Intractable Diseases
Dr. Hidehiro Mizusawa, Chairperson of the Prion Disease Surveillance Committee of the "Prion Disease Surveillance and Infection Prevention Research Group" funded by the Ministry of Health, Labour and Welfare of Japan

Please check one or the other for the following patients.

□ For both surveillance registry studies and natural history studies
□For the surveillance registry study only

I have obtained consent to cooperate in both the surveillance registry study and the natural history research study.
Please inform the secretariat of the telephone number and e-mail address of the patient or the surrogate obtained at the time of consent to participate in the natural history research, together with the consent document [Form 5] regarding the provision of the telephone number address, etc.

Date of birth

Name of attending physician:

Affiliation:

Name of attending physician

Patient's name (initials) Hepburn style (first name, last name)

Date of birth: (year, month, day)

Gender: Male / Female
(Circle one or the other)

Consenting party: Patient ・ Consenting party (Relationship to patient:)

Request to the attending physician
Please keep the consent document [Form 1] at your side to protect the patient's personal information. To indicate that consent has been obtained, please fill out this confirmation form ([Form 2]), convert it to PDF format, and send it to the Surveillance Research Office (prion-ncnp@ncnp.go.jp).

front【Form 3】 【Doc 6-3】

Date：30 November 2017

Consent Withdrawal Form

Grant-in-Aid for Scientific Research on Intractable Diseases
Dr. Hidehiro Mizusawa, Chairperson, Prion Disease Surveillance Committee, "Surveillance and Prevention of Prion Disease

National Surveillance Study on Prion Disease Surveillance and Natural History
Study 1 (Prion disease surveillance study (field survey))
Study 2 (JACOP natural history study)

I am.
□National Surveillance Study on Prion Disease Surveillance and Natural History
□Surveillance study of prion diseases
□I have given my consent to participate in the □National Prion Disease Surveillance and Natural History Study (please check all that apply), but have decided to withdraw that consent and submit a withdrawal of consent form.

Date of withdrawal Date of withdrawal

Signature

Address

Phone Number

Name of Alternate Signer/Substitute (if you are a minor/if it is difficult for you to give your consent with full understanding)
I,
_____
_____, hereby withdraw my consent to the following
□National Surveillance Study of Prion Disease Surveillance and Natural History
□National Prion Disease Surveillance Study
□I hereby withdraw my consent to participate in the National Prion Disease Surveillance and Natural History Study (please check all that apply) and submit the withdrawal of consent form.
Date of withdrawal Date of withdrawal

Signature (signature) (Relationship)

Address

Telephone number * In principle, I can withdraw my consent by submitting a withdrawal of consent form.

* In principle, the person who signed the consent form should be the one to withdraw consent.

back【Form 3】 【Doc 6-3】

I hereby certify that I have received the Consent Withdrawal Form as follows Date of Receipt Month Day
Recipient Affiliation

Signature of Recipient

Request to the attending physician
Please keep the Consent Withdrawal Form ([Form 3]) at your side to protect the patient's personal information. Also, please provide a copy to the patient. To indicate that consent has been withdrawn, please fill in the Consent Withdrawal Confirmation Form ([Form 4]), convert it to PDF format, and send it to the Surveillance Research Office (prion-ncnp@ncnp.go.jp).

【Form 4】 【Doc 6-4】

Prion Disease Surveillance and Prevention Group Prion Disease Surveillance (Field Survey) (or natural history survey)

Grant-in-Aid for Scientific Research on Intractable Diseases
Dr. Hidehiro Mizusawa, Chairperson of the Prion Disease Surveillance Committee of the "Prion Disease Surveillance and Infection Prevention Research Group" funded by the Ministry of Health, Labour and Welfare, Japan.

The following patients should be registered first.

□Patients who were included in both surveillance registry studies and natural history studies
□Surveillance registry study only
□For natural history studies only (Please check all that apply)

I wish to inform you that my consent to cooperate has been withdrawn.

Date of withdrawal

Name of attending physician: Affiliation

Attending physician's name: Attending physician's affiliation: Affiliation

Patient's name (initials) Hepburn style (first and last name):
Date of birth: (A.D.) Year Month Day Gender: Male / Female
(Please circle one of the following)

Consenting party: Patient him/herself・ Alternate consenting party (Relationship to the patient:)

Request to the attending physician
Please keep the [Form 3] Consent Withdrawal Form at your side to protect the patient's personal information.
To indicate that consent has been withdrawn, please fill out this confirmation form ([Form 4]), convert it to PDF format, and send it to the Surveillance Research Office (prion-ncnp@ncnp.go.jp).