Information for medical staff

Variant Creutzfeldt-Jakob disease (vCJD) and treatment with blood from 80 or more donors

This information is for clinicians and other staff caring for patients considered to be at risk of variant CJD (vCJD) for public health purposes following treatment with blood from 80 or more donors.

In this document, 'CJD' means all types of human prion disease, unless we say sporadic CJD or variant CJD.

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Health
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1 Advice for patients considered to be at risk of vCJD for public health purposes

If you are 'at risk of vCJD for public health purposes', you have been treated with blood or parts of blood, which came from 80 or more blood donors. You may have been treated with blood transfusions, cryoprecipitate, fresh frozen plasma (FFP) or platelets.

Although we believe the chances of your developing vCJD are very low, you should do the following things to reduce the chances of passing vCJD to other people.

- **Don't donate blood. You shouldn't do this anyway, because you have received blood donated in the UK since 1980.**
- **Don't donate organs or tissues.**
- **Tell whoever is treating you before you have any surgical, endoscopy or dental procedures, so they can make special arrangements for the instruments used in your care.**
- **It would be best if you tell your family about this in case you need health care in the future for any reason and your family can help by telling the healthcare staff.**

Tissue and organ donations

If you are at risk of vCJD for public-health purposes, you should not donate organs or tissues.

There is no evidence that vCJD can be passed from a woman to her unborn baby or by breastfeeding. To be cautious, if you are a man you should not donate sperm, and if you are a woman you should not donate eggs or breast milk.

Family, friends and others

You can't pass vCJD to your family, friends, healthcare workers or other people through normal contact. vCJD is not passed on by sneezing or coughing, by touching or by having sex. There is no evidence that vCJD could pass from a mother to her child.

1. Advice for health protection teams

Patients at risk of vCJD may have had surgery since the time when they were exposed to vCJD. If this is the case, instruments used on tissues with medium-low, medium or high infectivity levels, could infect other patients (see Table 1). This is because infectious prion proteins on the surgical instruments are not completely removed by the usual decontamination processes.

Table 1 Tissue infectivity levels for patients with, or at risk of vCJD

Infectivity level	Tissues of patients with, or at risk of vCJD
High	Brain Spinal cord Posterior eye
Medium	Olfactory epithelium Lymphoid tissue
Medium–low	Anterior eye
Low or not detectable	Other

The risk of transmitting vCJD from one patient to another on surgical instruments will fall each time the instruments are used and decontaminated. After being used and decontaminated around ten times (20 times for high-risk tissue), the instruments are not likely to infect other patients.

Health protection teams should report to the CJD Incidents Panel any surgery carried out on medium-low and medium-risk tissues involving instruments that may have been used and decontaminated less than ten times (20 times for high-risk tissues) since being used on a patient at risk of vCJD. Please use the reporting form available on the HPA website at www.hpa.org.uk/infections/topics_az/CJD/incidents_panel.htm.

Surgical departments reuse instruments at different rates. Reviewing the medical history over the last 12 months for each patient who is at risk of vCJD for public-health purposes should reveal any instruments that could still infect other patients.

You should also review the medical records of patients at risk of vCJD who have died in the last year.

The CJD Incidents Panel may decide that the instruments used on a patient at risk of vCJD should be quarantined immediately or destroyed. The Panel currently advises that there is no need to contact the patients on whom these instruments were used. However, the Panel will review this advice in line with any relevant new scientific evidence.

There is no need to investigate or report other operations that have been done on patients at risk of vCJD, as long as standard processes for decontaminating the surgical instruments have been used.

Please also report to the Panel any donations of blood, tissues or organs made by these patients since they became at risk of vCJD. The Panel advises that there is no need to contact patients who have received blood, tissues or organs donated by someone at risk of vCJD following treatment with plasma products. The Panel will review this advice in line with any relevant new scientific evidence.

2 Infection control advice

If you are carrying out an invasive procedure, operation, or are part of an infection control team, please follow the guide to controlling infections published by the Advisory Committee on Dangerous Pathogens Transmissible Spongiform Encephalopathy (ACDP TSE) Working Group in 2003. You can find the guide at www.advisorybodies.doh.gov.uk/acdp/tseguidance/Index.htm. This tells you what action you should take to control vCJD infection in hospital, during surgery, and in community health care such as dentistry.

If you are a patient who is 'at risk' of vCJD and you need to have an invasive medical procedure, you should tell the healthcare staff in charge of your care about this so they can take action to stop vCJD being passed on to other people.

Infection control abroad

If you are at risk of vCJD and you need an operation or endoscopy while you are abroad, you should do the following things to reduce the chances of vCJD being passed on to other people.

- Tell the medical staff caring for you that, in the UK, doctors must take special infection control measures when you have surgery, to reduce the risk of infecting other patients with vCJD.
- Tell the medical staff that they should contact their own national organisation for advice on controlling infections, who can then contact the Health Protection Agency (HPA) duty doctor on 0044 208 200 6868 to get advice about the safety measures recommended for the medical procedure they are planning to do, or have already done.

The duty doctors are available 24 hours a day every day, and have information about:

- where to find the relevant guidelines for controlling infection; and
- contact numbers for experts who can help interpret the guidelines if necessary.

If it is not possible to tell medical staff before they operate or give you treatment, you should tell them as soon as possible afterwards. This may happen in the UK as well as abroad, for example, if you are unconscious and need treatment straight away. Procedures for controlling infection can still be effective, even if you tell the healthcare staff after your operation (or other medical procedure).

3 Advice for GPs and other medical staff

If you are responsible for telling a patient they are at risk of vCJD for public health purposes, you should make sure that the patient's GP is informed.

The GP should do the following things:

- Know that their patient is being told they are at risk of vCJD for public-health purposes.
- In the patient's primary-care records, record that the patient is at risk of vCJD for public-health purposes and that special infection control measures are needed.
- Include this information in any referral letters if the patient needs surgery, an invasive procedure or dental surgery. Remember to inform consultants who are providing ongoing care to patients at risk of vCJD.
- Check if the patient has had surgery in the past year. If this is the case, the GP should tell the local health protection team, who will take action if necessary.

The doctor caring for patients who are at risk of vCJD should make sure that their 'at risk' status is recorded in their hospital medical records and primary-care notes. **This should only be done once the patient knows that they are at risk of vCJD.**

Doctors should ensure that patients at risk of vCJD receive the same medical and dental care that they would if they were not at risk.

Advice for dentists

If you have a patient who is at risk of vCJD, they should tell you about this. This will allow you to follow satisfactory standards for infection control. You should include the information if you refer your patient to specialists such as maxillofacial surgeons.

There is no reason why you should refuse dental care to any patients with, or at risk of, vCJD, or their relatives. You can treat them in the same way as any other patient.

In February 2005, the Chief Dental Officer sent a letter to all dentists in England to give information and advice about treating patients with (or at risk of) vCJD. You can find this letter at http://www.dh.gov.uk/en/Publicationsandstatistics/Lettersandcirculars/Dearcolleagueletters/DH_4102752

The CJD Incidents Panel and HPA have prepared an advice note concerning the provision of dental care to individuals 'at-risk' of CJD which can be found at http://www.hpa.org.uk/infections/topics_az/cjd/guidance.htm
The TSE guidance for controlling infection states that 'the risks of transmission of infection from dental instruments are thought to be very low, provided optimal standards of infection control and decontamination are maintained.'

You can deal with dental instruments you have used on patients with (or at risk of) vCJD in the same way as in any other low-risk surgery (that is, you

can decontaminate them in line with best practice and use them again) with the exception of endodontic instruments. In April 2007, the Chief Dental Officer issued a letter, Advice for dentists on re-use of endodontic instruments and vCJD which can be found at http://www.dh.gov.uk/en/Publicationsandstatistics/Lettersandcirculars/Dearcolleagueletters/DH_074001. This advises that endodontic reamers and files should be used only once. Also, you should never use any instruments that are labelled 'single use' more than once. This letter also contains advice on decontaminating dental instruments.

Discussing vCJD risks with patients

Patients may find the news that they are at risk of vCJD distressing. They may want an absolute guarantee that they will not develop vCJD. This is clearly not possible.

Variant CJD (vCJD)

Many people in the UK will have eaten food that could have been infected with BSE, which causes vCJD. We do not know how large the extra risk of developing vCJD is from treatment with blood from a large number of donors, but it is likely to be very low.

Four patients have been infected with vCJD following blood transfusions in the UK since 1980(?). One of these patients did not develop clinical disease.

As far as we know, vCJD has never been spread through surgery.

Sporadic CJD and other types of CJD

Everyone has a very small risk of developing sporadic CJD. We do not know how large the extra risk of developing CJD is from being operated on with instruments used before on someone with sporadic CJD, but it is likely to be very low.

In the whole world, there have been four cases of people developing CJD after being operated on with instruments used before for neurosurgery (surgery to the brain and spinal cord) on patients with sporadic CJD. These people developed CJD within about 18 months of being infected. Two cases have been linked to using infected depth electrodes on the brain and another two to corneal transplants.

Before informing at-risk patients, you should consider their risks carefully and make such preparations as giving them the opportunity to discuss these issues with appropriate healthcare staff afterwards.

You will need to decide who should be involved in telling patients about their risk of developing vCJD, and how this should best be done. Many patients are likely to need more than one session to discuss what this means for them if they are to come to terms with what they have been told. You may need to consult a trained counsellor for advice on managing this process.

Annex 1 How and why patients are identified as at risk of CJD for public-health purposes

The CJD Incidents Panel

CJD incidents happen when there is a possibility that patients could have come into contact with CJD through:

- treatment with plasma products such as clotting factors and albumin
- receiving a blood transfusion
- surgical instruments contaminated with CJD
- organ or tissue transplants.

In 2000 the UK Chief Medical Officers set up an independent committee of experts, the CJD Incidents Panel (the panel). This panel gives advice to all organisations responsible for providing health care on how to manage situations when CJD could have been passed on from one patient to another. In particular, the panel gives advice to local infectioncontrol teams and health protection teams.

Medical teams should ask for panel advice when they are dealing with patients who might have come into contact with CJD. The panel assesses the risk to these patients, and advises whether the team should contact the patients and tell them that they are at risk of CJD. Teams will then need to tell these patients about the safety measures that are necessary to prevent CJD spreading to others.

You can get more information on the panel from our website at www.hpa.org.uk/infections/topics_az/cjd/incidents_panel.htm. This includes the panel framework document, which sets out the principles of managing incidents where a patient may have come into contact with CJD. Our CJD section provides the secretariat for the CJD Incidents Panel, and provides help to healthcare teams that are following the panel's advice.

The panel's recommendations

The CJD Incidents Panel advises on the need to contact patients who have a risk of at least 1% of being infected with CJD through medical procedures such as treatment with plasma products. The patients should be told they are at risk of CJD for public health purposes, and that they should take certain safety measures to prevent CJD being spread to other patients.

Much of the scientific data that underpins the vCJD risk assessment models is uncertain. Where there has been doubt, the panel has made cautious assumptions. The risk assessment and 1% threshold provide a guide for deciding when safety measures are needed to limit CJD spreading from person to person. They should not be used as a precise guide for telling people their exact risk of developing CJD.

CJD risk from surgical instruments

Surgical (or other) instruments may be contaminated with prion protein (the protein that carries CJD) when they touch infectious tissues of a patient with CJD. Prion proteins are not completely destroyed in normal decontamination processes, and the instruments may spread CJD to other patients when they are used again.

The amount of material on an instrument after surgery depends on the type of instrument, the tissues it was used on, and the decontamination processes the instrument has been through. Usually, there are two stages for decontaminating surgical instruments. First, they are cleaned, for example in a mechanical washer and drier. Then, infectious material is inactivated, for example, by high-pressure steam autoclaving.

Studies show that the first cycle of decontamination may reduce the amount of protein on instruments with flat surfaces by a thousand-fold. However, instruments with serrated edges and hinges, or with narrow lumens (for example, flexible endoscopes) are much more difficult to clean. Cleaning the instruments again is not likely to be as effective, as any material that has survived the first cleaning stage may have been baked on during autoclaving or, in the case of flexible endoscopes, fixed to the equipment by the chemicals used in processing. It is possible that even after using and decontaminating the instruments a great many times, some infectious prion proteins could remain on the instruments.

The Department of Health has modelled the infection risk to patients coming into contact with surgical instruments that were used on a patient with CJD. These assessments show that the risk to patients is highest if the surgical instruments used on them have only been used and decontaminated a small number of times since being used on a patient with CJD. You can find this risk assessment on our website.

The panel's advice on whether to contact patients in an incident is based on these models. The panel assesses the risk of patients coming into contact with CJD by reviewing the information collected by the local incident team. In each case the panel considers a range of factors including:

- the clinical condition of the patient with CJD
- the infectivity levels of the tissues that the instruments have been used on
- the type of instruments used
- the processes in place for decontaminating the surgical instruments, and
- whether the instruments can be traced.

Annex 2 More information about CJD

Creutzfeldt-Jakob disease (CJD) is one of a rare group of diseases, known as 'transmissible spongiform encephalopathies' (TSEs), that affect the structure of the brain. TSEs cause dementia and a range of neurological symptoms, including ataxia and jerky movements.

A number of TSEs are recognised in both humans and other animals. In animals, the best-known TSE is bovine spongiform encephalopathy (BSE or mad cow disease). In humans, there are four main types of CJD:

- sporadic CJD, which accounts for 85% of cases
- variant CJD (vCJD)
- inherited CJD
- iatrogenic CJD (through medical procedures).

At the moment, we can diagnose CJD by histological examination of the brain, following a brain biopsy or after a post-mortem. If someone has symptoms suggestive of variant CJD, tonsil biopsies may also be used. There is no proven treatment or cure for CJD, and the disease always leads to death.

Research is being done on the causes, tests and possible treatments for the disease. One trial of a treatment has taken place at the NHS National Prion Clinic (NPC) and the Medical Research Council's (MRC's) Prion and Clinical Trials Unit.

Sporadic CJD

Sporadic CJD is most common in people over 50, and affects about one in a million people in the world. It is thought to arise spontaneously. Early symptoms are usually of behavioural disturbance or mental deterioration. A rapidly progressive dementia with obvious multifocal neurological involvement soon develops. Within weeks the patient may become unsteady on their feet, lack co-ordination and become very clumsy. In some people these are the first symptoms. Later symptoms may include blurred vision or even blindness, rigidity in the limbs, sudden jerky movements, and incontinence. Death usually occurs within months of the symptoms starting.

Variant CJD (vCJD)

Variant CJD was first recognised in 1996 and is thought to be caused by eating beef from cattle infected with BSE, although no-one knows the exact cause of infection. It usually affects younger people, with a median age of onset in the late 20s. The clinical picture is different from sporadic CJD in that it often starts with psychiatric symptoms, such as anxiety and depression. There may be persistent pain, with odd sensations in the face and limbs. These symptoms are followed by more obvious neurological symptoms and progressive dementia . Variant CJD is also different from other human TSEs because infectious prion proteins are found outside the nervous system as

well as inside, especially in the lymphoid tissues throughout the body. People with variant CJD tend to live longer than people with most other forms of CJD, with an average of 14 months between symptoms starting and death.

Over 150 people have died from vCJD in the UK and a small number have died in other countries. The number of cases in the UK is now declining and only five deaths were reported in the UK in 2006. The latest estimates are lower than some of the pessimistic forecasts that were made in the mid-1990s. However, nobody knows how many people will get this disease in the future. More information, including monthly numbers of cases and the latest short-term incidence projections are available from the National CJD Surveillance Unit's website, www.cjd.ed.ac.uk.

Inherited CJD

Inherited CJD has an autosomal dominant inheritance. The patients are often younger and live longer than people who develop sporadic CJD. There are between six and ten new cases each year in the UK. The clinical features of inherited CJD vary from person to person, even within one family. Some patients have signs and symptoms similar to those seen in sporadic CJD, while others develop ataxia and other movement disorders before dementia starts.

Iatrogenic CJD and CJD transmission through blood

People may develop iatrogenic CJD after infectious tissue enters their body through:

- surgery and other invasive medical procedures
- receiving infectious material (such as blood components), or
- having treatment with human hormones derived from the pituitary gland such as human growth hormone.

The clinical features of iatrogenic CJD partly depend on the route of infection. About 150 people have been infected after having received contaminated dura mater grafts. Over 100 people have been infected through treatment with contaminated human growth hormone.

Infection with variant CJD (vCJD) has probably been transmitted to three patients through blood transfusions in the UK. One of these patients did not develop clinical disease. No cases have been reported among patients who received plasma products sourced from people who went on to develop vCJD.

There is no epidemiological evidence that blood transfusions from donors who later develop sporadic CJD, can infect people. However, experiments in which blood from humans with sporadic CJD is injected into the brains of animals, suggest that blood might be infectious at a relatively low level. It is possible that some cases could have occurred, without this source being recognised.

How CJD spreads

Prion diseases like CJD can spread from one person to another in certain circumstances, but they are not infectious in the usual way. They are not spread by coughing or sneezing, touching or by having sex, nor is there evidence that the disease can spread from a mother to her unborn baby or spread through breastfeeding.

Eating beef or other products from BSE-infected cattle is the most likely cause of vCJD, and most of the people in the UK would have been exposed in this way. Other sources of vCJD infection include inoculation from contaminated medical equipment or receiving infected transplant material.

Preventing CJD from spreading between patients

The following public health measures aim to reduce as far as possible the chances of spreading CJD between people:

- improving the standards for decontaminating instruments
- taking special infection control measures when operating on patients with, or at risk of, CJD
- measures to protect the blood supply (see below).

Measures to protect the blood supply

Sporadic and inherited CJD

There have been no reports of CJD being spread by blood transfusions from a patient with sporadic or inherited CJD. Sporadic CJD has been monitored for many years in many countries, and this shows that transmission through blood is unlikely.

Variant CJD (vCJD)

We do not know the exact risk of vCJD spreading through blood. The Department of Health in England arranged for Det Norske Verita Consulting to assess this risk. The Spongiform Encephalopathy Advisory Committee (SEAC) accepted the risk assessment in early 1999.

As a result, the blood services take the following safety measures to reduce any possible risk of spreading vCJD through blood:

- withdrawal and recall of any blood components, plasma products or tissues donated by anyone who later develops vCJD (since December 1997)
- importing plasma from the USA for fractionation to make plasma products (since 1998)
- removing white blood cells (which may carry the highest risk of spreading vCJD) from all blood used for transfusions (leucodepletion) (since October 1999)
- importing fresh frozen plasma from the USA for patients born on or after 1^{t} January 1996 (since August 2002)

- not accepting (since April 2004) donations from people who have received a blood transfusion in the UK since 1980. In August 2004, this was extended to include people who are not sure if they have had a blood transfusion, and apheresis donors
- promoting the appropriate use of blood, tissues and alternatives throughout the NHS.

What is an infectious prion protein?

The cause of CJD is thought to be an abnormal form of the naturally-occurring prion protein. The normal form of this protein is found in the brain and other parts of the body, in humans and many other animal species, but we know little about its function. The abnormal infectious prion protein is chemically identical to the normal form, but its physical shape is different, and it resists normal cell degradation. We think it builds up by inducing the normal prion protein to mis-fold, but we do not know how this change happens. The abnormal prion protein then builds up in various tissues, particularly in the central nervous system, where tissue damage is most severe. As the disease progresses, neuronal tissue is lost, and the brain becomes 'spongiform'.

The abnormal prion protein that starts these changes may arise:

- spontaneously (a possible explanation for sporadic CJD)
- associated with an inherited abnormality of the prion protein gene (inherited CJD)
- acquired in a medical setting from contamination with tissue from someone with CJD (iatrogenic CJD), or
- acquired most likely through eating meat from cattle infected with BSE (for variant CJD only).

The immune system does not seem to respond to CJD infection. Also, the infected prion protein resists most of the usual methods to inactivate bacteria and viruses. Prions are not totally inactivated by heat, ultraviolet light or other standard sterilisation procedures such as immersion with sodium hypochlorite at normal concentrations. We cannot rely on autoclaving to denature abnormal prion proteins contaminating surgical instruments following use on a patient with CJD.

Of those who have been tested, most people with sporadic CJD, and everyone with variant CJD, have a particular form of the prion protein gene that is found in 40% of people in the UK. This genotype probably makes the prion protein more vulnerable to conversion into the abnormal form. In a post mortem carried out in July 2004, variant CJD infection was detected in the spleen and one cervical lymph node of someone who had a different form of the prion protein gene. The patient had died from a cause unrelated to vCJD. This happened some years after receiving a blood transfusion from a donor who later developed vCJD. The patient had not become ill with vCJD and we do not know if they would ever have done so.

Annex 3 Useful websites

You can get more information about CJD from the following websites and phone numbers.

Health Protection Agency
Website: www.hpa.org.uk/infections/topics_az/cjd/menu.htm

CJD Incidents Panel

Website: www.hpa.org.uk/infections/topics_az/cjd/incidents_panel.htm

Infection control guidance
Website: www.advisorybodies.doh.gov.uk/acdp/tseguidance/

Department of Health information for dentists
Website:

www.dh.gov.uk/PublicationsAndStatistics/LettersAndCirculars/DearColleagueLetters/DearColleagueLettersArticle/fs/en?CONTENT_ID=4102752&chk=7HspA

National CJD Surveillance Unit
Website: www.cjd.ed.ac.uk/index.htm

National Prion Clinic
Website: http://www.nationalprionclinic.org/
Phone: 020 7837 3611

Patient support groups

CJD Support Network
Website: www.cjdsupport.net/
Phone: 01630 673973

Human BSE Foundation
Website: www.hbsef.org/

CJD Therapy Advisory Group guidance:
Website:
www.dh.gov.uk/PolicyAndGuidance/HealthAndSocialCareTopics/CJD/CJDGeneralInformation/CJDGeneralArticle/fs/en?CONTENT_ID=4032403&chk=LVJY6b

Department of Health
Website: www.doh.gov.uk/cjd/index.htm

National Public Health Service for Wales
Website: www.wales.nhs.uk/sites/home.cfm?OrgID=368

Spongiform Encephalopathy Advisory Committee
Website: www.seac.gov.uk/

Department for Environment, Food and Rural Affairs BSE home page
Website: www.defra.gov.uk/animalh/bse/index.html

We last revised this leaflet in XXXXXX 2007. To check for any updates to this information, please see the current version of this leaflet, 'Variant Creutzfeldt-Jacob Disease (vCJD) and highly transfused – clinical information'. You can find this document at www.hpa.org.uk/infections/topics_az/cjd/information_documents.htm
We welcome your comments on this leaflet. Please send them to cjd@hpa.org.uk