Health Protection Agency

Scottish Centre for Infection and Environmental Health

NHS National Services Scotland

National Public Health Service for Wales
Gwasanaeth Iechyd Cyhoeddus Cenedlaethol Cymru

Department of Health, Social Services and Public Safety
An Roinn Sláinte, Seirbhísí Sóisialta agus Sábháilteachta Poiblí

VARIANT CREUTZFELDT-JAKOB DISEASE and PLASMA PRODUCTS

INFORMATION FOR PATIENTS

1. What is variant Creutzfeldt-Jakob disease?

Creutzfeldt-Jakob Disease, or CJD, is one of a group of rare and fatal diseases in humans and animals that affect the structure of the brain.

There are four main types of CJD: of these, sporadic CJD (arising spontaneously) is the most common and accounts for 85% of cases. The other types are familial, iatrogenic (through medical treatment) and variant CJD (vCJD). In animals the best-known TSE is bovine spongiform encephalopathy (BSE or 'mad-cow disease'). Variant CJD is believed to be the human form of BSE.

Many people living in the UK have been exposed to BSE (Bovine Spongiform Encephalopathy or 'mad-cow disease') from eating infected beef and so are at a possible risk of developing vCJD.

2. What's this about?

Late last year the death of a person from vCJD who died some years after receiving a blood transfusion from a donor who themselves died of vCJD, was announced. This was the first case of transfusion-associated vCJD infection and increased concern about the possible infectivity of blood. A second probable case was reported in July 2004.

When a patient is diagnosed with vCJD, the UK Blood Services are informed and checks are made to find out whether the patient ever donated blood. Blood products include blood components, which are derived from a single donation of blood, or pools of up to six donations; and plasma products, which are prepared from the pooled plasma of several thousand blood donations in a process known as 'fractionation'.

To date, nine people are known to have donated blood before they became ill with vCJD, and their donations were used to make plasma products. Thus a number of patients may have been exposed to vCJD infection in the course of their past medical care.

This information sheet about vCJD has been developed with doctors' and patients' groups for patients who have been informed by their doctor that they are considered 'at-risk' We hope it will go some way to answering your first questions.

3. Why am I being contacted?

Patients who have received implicated plasma products may be at an additional risk of vCJD. This risk is 'additional' since it is on top of the general risk for many people in the UK from eating beef in the past.

It is impossible to put an exact figure on the chances of getting vCJD, either from BSE-infected beef and beef products, or the possible additional risk from receiving implicated plasma products. So far there have been no cases of vCJD amongst recipients of plasma products sourced from blood donors who later developed vCJD, and the risk of this happening is likely to be very low.

If you received implicated plasma products there is a small possibility that vCJD could have been passed on to you. If so it might be possible for you to pass vCJD on to others in certain circumstances, in which case you and the people providing your healthcare need to take some special precautions to avoid putting other people at potential risk. This is why it is important that you know, even if this causes you anxiety.

4. What measures are already being taken for vCJD?

A number of measures have minimised the risk of getting vCJD from eating BSE-infected meat and meat products. These include banning the feeding of animal protein to other animals, and removing certain parts of animals from the food we eat.

In the healthcare setting, the abnormal 'prion' protein, the infective agent that causes vCJD, is very hard to destroy. Using surgical instruments only once, or destroying those that have been used on patients diagnosed with vCJD, is one way to guard against passing on vCJD. In recent years much effort has also gone into ensuring that the decontamination of all surgical instruments is to the highest standards. The aim is to remove as much potentially infected material as possible.

5. And in relation to blood?

Because it is uncertain whether vCJD can be transmitted by blood the United Kingdom blood services have taken a number of precautionary measures:

- Withdrawal and recall of any blood components, plasma products or tissues obtained from any individual who later developed vCJD (December 1997),
- Importing plasma from the US to manufacture plasma products (1998),
- Removal of white blood cells (leucodepletion) from all blood components (Autumn 1999),
- Importing fresh frozen plasma from the US for patients born on or after 1st January 1996 (Autumn 2002),
- Not accepting donations from people who have received a blood transfusion since 1980 (April 2004),
- Promoting appropriate use of blood and tissues and alternatives throughout the NHS.

6. And in relation to patients?

In 2000 an expert advisory committee called the CJD Incidents Panel (CJDIP) was set up to advise on the handling of 'incidents' of possible transmission of CJD, including vCJD, in a healthcare setting. The CJDIP assesses the risk to other patients, and advises whether patients should be contacted and informed about their possible exposure.

The CJDIP has agreed that, in general, where patients may have been exposed to a 1% or greater possible risk of infection[1], they may have an additional unknown risk of developing vCJD, on top of the general risk from eating beef in the past, and should be contacted. These patients should be given advice about what they should do to avoid putting other people at risk. This advice includes not donating blood, tissue and organs, and informing healthcare professionals so that extra precautions can be taken if they require invasive medical or dental procedures, for example a surgical operation.

There are a lot of uncertainties in estimating the risk of infection with vCJD and a very cautious approach has been taken. The CJDIP has chosen this 1% threshold for informing patients of their exposure so that special precautions can be taken to limit the possible risk of transmitting vCJD between patients. This is considered the best balance between protecting the public from further spread of vCJD and causing excessive anxiety regarding a risk which is uncertain, but thought to be low.

7. So what's new?

Since the CJDIP was established it has been considering policy towards recipients of blood from donors who later developed vCJD.

When people are diagnosed with vCJD, any blood donations they have given are traced. The CJDIP has estimated the potential additional risk of vCJD from treatment with plasma products sourced from all donors known to have later developed vCJD. This risk depends on the type of plasma product and how each batch was manufactured, as well as the amount a patient may have received.

For certain plasma products (e.g. intramuscular immunoglobulin used for travel vaccinations against hepatitis A, or anti D for Rhesus negative pregnant women) the amount of estimated infectivity in the implicated products is so low that the possibility of reaching the 1% threshold can realistically be ignored. Patients who have received these products do not need to take any special precautions.

For other products (e.g. clotting factors and antithrombin, intravenous immunoglobulin, albumin 4.5%) the infectivity may be higher, depending on how the product was made. Once one of these plasma products has been identified the next step is to try to identify those patients who are likely to have had sufficient product to reach the 1% threshold and who need to take special precautions. These patients are considered to be 'at-risk' of vCJD for public health purposes.

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[1] A 1% risk of infection means that there is a 1 in 100 possibility that vCJD can be transmitted.

8. Who is affected?

Patients who are considered 'at-risk' of vCJD for public health purposes will be informed by their doctor. The people who may be affected are in three main groups:

- some patients with bleeding disorders (including congenital and acquired haemophilia (haemophilia A and haemophilia B), Von Willebrand Disease, other congenital bleeding disorders) and congenital antithrombin III deficiency,
- some patients with primary immunodeficiency (PID), and
- some patients with other illnesses who might be considered 'at-risk'. These may include, patients with secondary immunodeficiencies; certain neurological conditions and autoimmune illnesses (such as idiopathic thrombocytopaenic purpura), plasma exchange recipients and patients with severe burns. Patients with certain other conditions requiring critical care (including acquired antithrombin deficiency or patients requiring rapid warfarin reversal) may also be affected.

9. How does this affect me?

If you have been informed that you are 'at-risk' for public health purposes, you are being asked to take the following actions in order to reduce the chance of passing on vCJD to other people:

- **Do not donate blood.**
- **Do not donate organs or tissues.**
- **Tell whoever is treating you before you undergo medical, surgical or dental treatment, so they can then arrange any special procedures for the instruments used in your care.**
- **It would be best if you tell your family about this in case you might need emergency surgery in the future.**

A note of this will be made in your hospital medical records and your GP notes. Your care should not be compromised in any way – it will be just the surgical instruments that will be treated differently. Nor will you need extra medical follow-ups because you are 'at-risk' for public health purposes. However, your doctor will always be willing to see you if you have any worries about your health.

10. So if I'm 'at-risk' for public health purposes - what happens now?

You need do nothing other than follow the advice given above (see Section 9).

Normal social contact and household activities do not spread the infection. Your family and friends are not at risk from you and you do not need to take any special precautions in your normal life.

Variant CJD is not infectious in the usual ways. There is no evidence that it can be passed on between people by sneezing or coughing (like colds and flu), sharing utensils, by skin contact, or through kissing or sexual intercourse.

There is also no evidence that vCJD can be sexually transmitted or transmitted from parent to child. However, as a precautionary measure, men who are 'at-risk' of vCJD for public health purposes should also not donate sperm.

11. Does this mean I'm going to suffer from vCJD?

Having reached the 1% threshold does not mean you will actually develop vCJD. This risk is unknown, but the chances of it happening are very low.

There is no evidence for transmission of vCJD by plasma products. Although the process of estimating risk is based on the best evidence available, there is much uncertainty about many aspects. As a result a cautious approach has been taken and may have over-estimated the potential additional risk of vCJD from receiving the various implicated plasma products. Despite these limitations it is still important to take extra public health precautions to provide the best protection for the population in general.

12. Can I be tested to see if I am infected?

No. Scientists are working very hard to develop a test, but as yet there is no test available that can be used to identify someone who may have been infected. Variant CJD can only be reliably diagnosed by brain biopsy or through examining the body after death.

13. What happens if I develop strange symptoms?

CJD causes dementia and a range of other symptoms, including difficulty with balance and extreme clumsiness. Unlike the other forms of CJD, vCJD often starts with psychiatric symptoms like depression and anxiety.

Go and see your doctor. It is unlikely that 'strange symptoms' will be the start of vCJD but your doctor will be able to arrange for you to see an expert if appropriate.

14. Will this mean I won't be able to get life insurance?

The Association of British Insurers have informed the CJDIP that their members will not refuse insurance just because someone is categorised as 'at-risk' for public health purposes.

15. General information about vCJD

What is the cause of vCJD?

Infections like influenza and pneumonia are caused either by viruses or bacteria. Some stomach infections are caused by microscopic parasites. Variant CJD, and the other TSEs, are different from these common infections. The cause is an abnormal infectious protein known as a 'prion'.

There is no test, treatment or cure for vCJD at present and the disease is always fatal. Scientists are researching the causes and possible tests and treatments for the disease.

How do you catch vCJD?

Variant CJD is believed to be caused in the first instance by exposure to the abnormal prion protein that causes BSE. Many of the UK population have been exposed through eating BSE-infected beef and beef products in the 1980s and early 1990s.

Variant CJD may also be transmitted between patients in the healthcare setting. So far there are no recorded instances of vCJD being spread through surgery, nor have there been any cases amongst recipients of plasma products sourced from individuals who later developed vCJD.

How many cases of vCJD are there?

So far, almost 150 cases of vCJD have occurred in the UK and a handful in other, mainly European, countries.

It is thought that the UK epidemic may have reached a peak. However no one knows how many people will contract this disease in the future.

16. Sources for Additional information

The process of informing patients about their possible additional risk status, and the special precautions they may need to take is being coordinated by the Health Protection Agency (HPA) in England, Wales and Northern Ireland, and in Scotland by the Scottish Centre for Infection and Environmental Health (SCIEH).

More information about vCJD with useful links is available from their websites
HPA: http://www.hpa.org.uk/infections/topics_az/cjd/menu.htm
SCIEH: http://www.show.scot.nhs.uk/scieh

Further information is also available from:

The Haemophilia Society http://www.haemophilia.org.uk
The Primary Immunodeficiency Association http://www.pia.org.uk
CJD Support Network http://www.cjdsupport.net
Human BSE Foundation http://www.hbsef.org
National CJD Surveillance Unit http://www.cjd.ed.ac.uk
Department of Health http://www.doh.gov.uk/cjd/index.htm
National Prion Clinic
http://www.st-marys.org.uk/specialist/prion/index_prion.htm
National Public Health Service for Wales
http://www.wales.nhs.uk/sites/home.cfm?OrgID=368
NHS Direct Online http://www.nhsdirect.nhs.uk

NHS Direct and its national colleagues are also operating a 'vCJD and Plasma Products' advice line for general enquiries (telephone: 0845 850 9850).