Fact sheet: Variant CJD & Blood Transfusions

About the CJD Support Network

The CJD Support Network is the leading care and support charity for all forms of CJD. The CJD Support Network:

- Provides practical and emotional support to individuals, families and professionals concerned with all forms of CJD
- Provides emotional support to people who have been told that they are at a 'higher risk' of CJD through blood or surgical instruments
- Links families with similar experiences of all forms of CJD
- Offers financial support for families in need
- Provides accurate, unbiased and up-to-date information and advice about all forms of CJD
- Provides a national helpline on all forms of CJD
- Promotes research and the dissemination of research findings
- Promotes good quality care for people with all forms of CJD
- Encourages the development of a public policy response for all forms of CJD
- Provides support, education and training to professionals concerned with CJD

For more information about the activities of the CJD Support Network, contact:

Admin and general enquiries - admin@cjdsupport.net or +44 (0)7494 211476
Support - support@cjdsupport.net or 0800 774 7317
Website - www.cjdsupport.net
Post - PO Box 3936, Chester, CH1 9NG

Registered charity no. **1097173**

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Contents

Background

The four types of human prion disease

- Sporadic Creutzfeldt-Jakob Disease (CJD), which arises spontaneously, is the most common type.
- Genetic or inherited prion disease is a rare form of the disease which has genetic causes.
- Iatrogenic CJD is accidentally transmitted through medical procedures/treatments.
- Variant CJD (vCJD) arose from BSE dietary contamination.

Visit www.cjdsupport.net/what-is-cjd/fact-sheets to access individual fact sheets on each form of CJD.

Variant CJD (vCJD)

In the 1980s and 1990s, there was an epidemic of a prion disease in cattle (BSE) which particularly affected the UK. Various control measures were put in place to eliminate cattle BSE and to protect human diet. As of November 2021, there have been 178 cases of definite or probable vCJD identified in the UK, the last case was reported in 2016, and no-one born after 1989 has so far been diagnosed with vCJD.

vCJD and Secondary Transmission

Following the identification of vCJD as a primary transmission from cattle to man (through diet) there were concerns that secondary human-to-human transmission might occur through surgery or blood. It is known that, when they are acquired as infections, prion diseases generally have long incubation periods (ie there is a long interval, typically measured in years, between being infected and becoming ill). In the case of vCJD, there is silent infection in parts of the body (such as the appendix or spleen) before the brain is affected with resulting symptoms. As such individuals are well, there is no obvious evidence they harbour infection. At present, it is thought that up to around 1:2000 of the UK population may be silently infected, with the potential for secondary transmission via surgery or blood. In fact, no instance of vCJD transmission has been identified by any route other than intravenous blood transfusion and an injected blood-derived product ('Factor VIII').

There are 3 instances of people developing vCJD after transfusion with blood from donors who were in the incubation period of vCJD. In one further case, a blood recipient later died of a non-neurological illness and evidence was found of vCJD infection but not affecting the brain. In one case, someone with haemophilia died from a non-neurological illness also with evidence of vCJD infection, thought to be due to Factor VIII treatment (produced from blood from a donor in the incubation period of vCJD).

It should be stressed that:

- Such secondary transmissions have occurred rarely (on only these 5 occasions) and no further instances have been identified for some 15 years (as of November 2021).
- No instances of vCJD transmission via surgery (including dental surgery) have been identified.
- There is no risk of transmission through ordinary contact, including intimate contact, with someone who has vCJD or might be incubating it.

What has been done to protect people?

People who are ill with vCJD will not be accepted as donors (as is the case with other illnesses). However, there is no method to detect asymptomatic infection that might be present in a donor. Blood tests for vCJD have been developed but they have not been evaluated as screening tests for asymptomatic infection in donors. They are also small-scale laboratory tests that are not yet suitable for the routine large-scale testing a Blood Transfusion Service requires.

Various measures have been taken to reduce the risk from blood including leuco-reduction of donated blood (reducing the white blood cell content), and exclusion from donation for those who have received blood in the past or were donors to known vCJD patients.

However, blood (in the form of red blood cells) is necessary to replace blood loss during surgery or due to trauma and this cannot be imported from other countries. It is important to appreciate that red blood cell transfusion may be life-saving in many circumstances.

People informed they are at risk of developing vCJD because of blood

If someone is identified as having received blood from a donor who went on to develop vCJD they are informed that they are 'at risk' because of having been possibly exposed to infection in that blood. In addition, if someone donated blood to another person who later developed vCJD, the donor is informed of being 'at risk' because of the possibility that the recipient was infected by the blood (i.e. that the donor might be asymptomatically infected). Strictly speaking, such individuals are at a higher risk than the general population-there is a risk in the general population due to past potential dietary exposure to BSE.

What is the actual risk?

Unfortunately, we don't know the precise risk. The fact that no-one has died from blood transfusion transmitted vCJD for over 15 years is obviously reassuring, but scientists are not complacent and remain vigilant for new cases. Most people in the UK are at some risk of developing vCJD because they have eaten beef or beef products contaminated with BSE in the past (click **here** to visit our fact sheet on vCJD. Just because someone has received blood that potentially contained vCJD infectivity, it does not necessarily mean that the blood actually contained infectivity, and, even it did, that the recipient will be actually infected. Finally, it is not necessarily the case that someone infected by blood will develop vCJD illness. It is possible that the infection will remain 'asymptomatic' (silent) in some people and will not affect their health.

How soon would anyone know if they will develop vCJD?

Unfortunately, this is difficult to answer. Firstly, the incubation period (time from infection to developing illness) in prion diseases can be very long; it is generally measured in years, rather than in weeks or months). Secondly, since only 3 people have developed vCJD because of blood transfusion, there is very limited evidence about the incubation period related to blood infection. In these 3 individuals, vCJD symptoms developed at intervals of 6½ to just over 8 years.

Can I be tested for vCJD infection?

There is no simple test that has been fully validated to determine whether someone has been asymptomatically infected by a blood transfusion. If there were such a test, it would determine only that there was asymptomatic infection but not necessarily whether this had resulted from the blood transfusion or from dietary BSE exposure. It would also not determine whether the infection would remain asymptomatic or result in vCJD illness.

What should I do if I am notified of being 'at risk'?

There is no specific action you should take in your ordinary life. There is no evidence that vCJD infection can be transmitted by ordinary, even intimate, contact. However, as a public health measure, you will be informed that certain precautions will be taken if you require certain medical investigations or treatments, in order to prevent any possibility of transmission to others. You should not donate blood, tissue or organs. Also, before undergoing any medical, surgical or dental procedure, you should tell the person carrying out the treatment about your potential risk, so that they can decide if any special arrangements for instruments they will be using are needed. It should be stressed that any measures taken are essentially precautionary, relevant to all those 'at risk', and do not mean that you are actually infected or infective. However, as a person 'at risk for public health purposes',

Your medical and dental care should not be compromised in anyway. A note of your risk will be made in your medical notes and there may be some occasions where instruments used in your care are treated differently. The National Prion Clinic and NCJDRSU may be able to help you if you feel that your care has been compromised.

What are the symptoms?

All forms of CJD have a range of psychiatric and neurological symptoms owing to the changes that occur in the brain. These are detailed in other fact sheets (click **here** to view our vCJD factsheet). It is important to understand that the early symptoms of vCJD are non-specific i.e. they may occur for many reasons other than vCJD. If you are concerned about symptoms of any sort, it is unlikely that they are the start of vCJD. Make an appointment to see your GP, who will refer you to a specialist if appropriate. You may also contact the CJD support network for general information and advice.

Are there any treatments?

Unfortunately, there are no treatments for the various forms of CJD at the moment. However, researchers all over the world are working towards developing treatments for CJD.

Can I pass the disease on to my children or family?

There is no evidence that vCJD contracted through a blood transfusion can be passed on from patient to child. Should you develop vCJD with or without symptoms, your children and other relatives will not inherit the disease. However, as a precautionary measure, people at risk should not donate sperm or breast milk.

Can being identified as at risk affect my ability to get life insurance, a mortgage or private healthcare?

Being at risk should not affect your existing policies. However, when taking out a new policy, you should answer all questions truthfully because if you lie or conceal information your policy could be invalid. If you are not asked questions relevant to your at risk status you do not need to volunteer information.

The Association of British Insurers has stated that its members do not refuse insurance just because someone is considered at risk for public health purposes. Mortgage companies and private healthcare agencies should adopt a similar approach. However, the CJD support network knows of one case where someone at risk was refused life insurance. If you have any problems obtaining insurance or other financial arrangements, please contact the CJD Support Network helpline on **0800 774 7317**.

Should I tell my employer?

There is no need to tell your employer about being at risk unless you are asked directly by, for example, your occupational health department.There is no evidence that CJD can be transmitted in the workplace. There have been no cases where doctors or the healthcare workers have passed CJD on to patients or where patients have transmitted the disease to workers in the health service.

Will this increased risk affect my health care?

Routine health and dental care should not be affected. However, as a precaution, your doctor, dentist or healthcare worker needs to be aware of your health risk status in the unlikely event of you needing complicated surgery.

I'm still concerned, what should I do?

If you are concerned about your health you should contact your GP. If you and your family would like to discuss your increase risk status you can contact either the National Prion Clinic, the National CJD Research and Surveillance Unit, or the CJD Support Network for advice.

Where can I get further information and support?

The information and that you are at risk of CJD for public health purposes has probably come as a shock. You may feel isolated, bewildered and unsure of where to obtain further information and support. There are a number of agencies that can provide you with more detailed information. In particular we would encourage you to contact the CJD support network. The network offers practical and emotional support for those diagnosed with any form of CJD, those considered at risk of developing CJD, their friends and families.

Further information and contacts

Further information about CJD may be found on the CJD Support Network website at www.cjdsupport.net our fact sheets are also available by post on request to the Network.

Support and information may be obtained from the organisations below:

CJD Support Network

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National CJD Research and Surveillance Unit

Western General Hospital, Crewe Road, Edinburgh EH4 2XU
Website - www.cjd.ed.ac.uk
0131 537 1980 telephone number for general enquiries and loth.securecjd@nhslothian.scot.nhs.uk for Care Team enquiries/patient families

National Prion Clinic

National Prion Clinic, Institute of Prion Diseases, Courtauld Building, 33 Cleveland Street, London, W1W 7FF.
Helpline for National Prion Clinic - 020 7679 5142 / 020 7679 5036
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