Public Health England

Health Protection Scotland

Protecting and improving the nation's health

Who has an increased risk of Creutzfeldt-Jakob disease?

This leaflet explains why different groups of people have an increased risk of Creutzfeldt-Jakob disease (CJD). Please read this together with the leaflet 'Information for people with an increased risk of CJD'.

Several groups of people have an increased risk of CJD. Everyone in these groups should follow advice to reduce the risk of spreading the infection to other people through their medical care.

The following groups of people have an increased risk of CJD:

1. **Related to blood and plasma:**

- people who have received blood from a donor who went on to develop variant CJD
- people who have given blood to someone who went on to develop variant CJD
- people who have received blood from a donor who has also given blood to another person who went on to develop variant CJD
- people who have been treated with certain plasma products produced in the UK between 1990 and 2001
- people who have received blood from 300 or more donors since 1990

2. **Related to surgery:**

- people who have had surgery using instruments that have previously been used on someone who went on to develop CJD
- people who have had a neurosurgical procedure or an operation for a tumour or cyst of the spine before August 1992
- people who have received an organ or tissue from a donor infected with CJD or at increased risk of CJD

3. **Related to other medical care:**

- people who have been treated with growth hormone sourced from humans before 1985
- people who have been treated with gonadotrophin sourced from humans before 1973
- people who have been told by a specialist that they have a risk of developing an inherited (genetic) form of CJD or other human prion disease that runs in families

If you have been infected with CJD, then you could spread CJD infection to other patients if you donate blood, organs and tissue or have an operation. Please follow our advice to help reduce the risk of spreading the infection to other people. If you are going to have an operation, special precautions should be taken with the surgical instruments that are used on you. This should reduce the risk of CJD (including variant CJD) being passed to others in operations.

Advice on how to stop CJD spreading to other people

If you have been identified as being at increased risk of CJD the following advice can reduce the risk of spreading CJD to other people:

- don't donate blood. No-one who is at increased risk of CJD or who has received blood donated in the United Kingdom since 1980 should donate blood
- don't donate organs or tissues, including bone marrow, sperm, eggs or breast milk
- if you are going to have any medical or surgical procedures, tell whoever is treating you beforehand so they can make special arrangements for the instruments used to treat you
- tell your family about your increased risk. Your family can tell the people who are treating you about your risk of CJD if you need any medical or surgical procedures in the future and are unable to tell them yourself

The following section explains how an increased risk of CJD was identified and what we know about these different types of risk.

1. Related to blood and plasma

We know that variant CJD (vCJD) can be spread by blood transfusions. Four people in the UK have been infected in this way. Three of these people developed vCJD. The fourth person had no symptoms of vCJD and died of an unrelated cause. This person's infection was only detected at post-mortem. It is impossible to put an exact figure on your increased risk of vCJD and the chance that you are infected with vCJD is very small. However, there are no tests at present that can identify a person who is infected with vCJD before they become ill, or that can detect blood that is infected with vCJD. People who are infected with vCJD may remain well for many years, and it is possible that some may never become ill. Most people who have vCJD have acquired it through eating beef or beef products from cattle that were infected with bovine spongiform encephalitis (BSE).

1.1 People who have received blood from a donor who went on to develop variant CJD

Your medical records show that you have received blood from a donor who later developed variant CJD (vCJD). Everyone who received blood from donors who went on to develop vCJD has been informed that they may have been exposed to vCJD.

1.2 People who have given blood to someone who went on to develop variant CJD

Your medical records show that your blood was given to a patient who later developed variant CJD (vCJD). We cannot tell how this patient became infected with vCJD. Most people who have vCJD acquired it through eating beef and beef products from cattle that

were infected with BSE. It is possible that this patient's vCJD infection came from your blood and that you could be infected with vCJD, even though you feel healthy.

1.3 People who have received blood from a donor who has also given blood to someone who went on to develop variant CJD

Your medical records show that you have received blood from a donor who has also given blood to someone who later developed variant CJD (vCJD). We do not know how that person became infected with vCJD. Most people who have vCJD have acquired it through eating beef or beef products from cattle that were infected with BSE. It is possible that the blood donor was the source of that patient's vCJD infection. If so, it is also possible that you may have been infected with vCJD from the same blood donor. The blood donor has no signs of CJD, and no longer donates blood.

1.4 People who have received blood from 300 or more donors since 1990

Your medical records show that you have received blood, cryoprecipitate, fresh frozen plasma or platelets from 300 or more blood donors. The larger the number of people who have donated blood to you, the greater the chance that one of them was infected with variant CJD (vCJD). No one who has donated blood to you has become ill with vCJD. If this were to happen, we would contact you via your doctor. It is possible that one or more of the donors who gave blood to you was infected with vCJD even though they appeared to be healthy. If so, they could have passed the infection on to you through their blood.

1.5 People who have been treated with certain plasma products produced in the UK between 1990 and 2001

One haemophilia patient has been found to have evidence of variant CJD (vCJD) infection when tested at post-mortem. This patient did not have any symptoms of vCJD, and died of an unrelated cause. The most likely source of this patient's infection was UK sourced clotting factors.

1.6 People with bleeding disorders who have been treated with clotting factors or antithrombin

Your medical records show that you have a bleeding disorder or congenital antithrombin III deficiency[1] and you have been given clotting factors or antithrombin prepared from plasma (a part of blood) sourced and produced in the UK[2] between 1990 and 2001.

You have an increased risk of variant CJD (vCJD) because you are one of many patients treated with clotting factors prepared from the plasma of a great many donors. The larger the number of people that have donated plasma that was used to prepare these clotting factors, the greater the chance that one of them was infected with vCJD, even though they appeared to be healthy. We do not know your increased risk of vCJD from treatment with clotting

[1] Congenital and acquired haemophilia (Haemophilia A and Haemophilia B), Von Willebrand Disease, other congenital bleeding disorders and congenital antithrombin III deficiency.
[2] Factor VIII, factor IX, factor VII, factor XI, factor XIII and prothrombin complexes, as well as antithrombin.

factors or antithrombin. The chance that you are infected with vCJD is thought to be very small.

1.7 People treated with albumin, and people without a bleeding disorder who have been treated with Factor IX (eg to reverse anticoagulant therapy)

Assessment of your medical records shows that you have been treated with albumin or Factor IX made from plasma (a part of blood) that was donated by someone who went on to develop variant CJD. We do not know what the chance is that you have been infected with variant CJD from treatment with albumin or Factor IX but it is likely to be very small.

2. Related to surgery

2.1 People who have had an operation using instruments that have previously been used on someone who went on to develop CJD

Your medical records show that you have had an operation using surgical instruments that have previously been used on a patient who went on to develop CJD.

Surgical instruments used on patients who are infected with CJD could spread infection to other patients having surgery. This is because the abnormal prion proteins that cause CJD are very hard to destroy. Surgical instruments that have been properly washed and disinfected may still have infected prion proteins on them and could then spread CJD to other patients.

We do not know what the chance is of getting CJD after having surgery, but it seems to be very small. Worldwide, there have been four reports of people infected with sporadic CJD from instruments used during neurosurgical procedures on the brain and spinal cord, and two reports of people infected with sporadic CJD from electrodes used on the brain; the most recent case was more than 30 years ago. We do not know of any people infected with variant CJD as a result of surgery.

2.2 People who have had a neurosurgical procedure, or an operation for a tumour or cyst of the spine before August 1992, and who may have received a graft of dura mater tissue in this procedure

Your medical records show that you had a neurosurgical procedure before 1992. You may have received a graft of dura mater tissue from humans during this procedure. Dura mater is a tough lining round the brain and spinal cord. Dura mater grafts obtained from humans used to be used in many neurosurgical procedures and operations on tumours or cysts of the spine. Many thousands of people have been treated with these grafts throughout the world. More than 220 of these patients worldwide have developed CJD, and the grafts that they received must have been made from people who had been infected with CJD, even if they had shown no signs of the disease while they were alive. These grafts were banned in the UK in 1992.

Everyone who has received a dura mater graft obtained from humans has an increased risk of CJD. If your operation took place some years ago, the hospital may no longer have a record of whether you received a graft of dura mater tissue from humans, and you should follow the same advice as someone who knows that they were given a graft of human tissue.

2.3 People who have received an organ or tissue from a donor infected with or at increased risk of CJD

Your medical records show that you have received an organ or tissue donated by someone who is at increased risk of CJD.

We do not know what your chance is of getting CJD after receiving an organ or tissue transplant but it is very likely that the benefits of your transplant are much greater than the risk of being infected with CJD. You may wish to discuss this with your surgeon.

We do not know of anyone who has been infected with variant CJD from an organ or tissue donation. Two people are thought to have been infected with CJD by corneal grafts donated by people who had sporadic CJD. These donations took place in the 1960s and 1970s.

3. Related to other medical care

3.1 People who have been treated with growth hormone from humans before 1985

Your medical records show that you were treated with growth hormone prepared from human pituitary glands between 1958 and 1985. Many thousands of children were given this treatment, and over 220 are known to have developed CJD worldwide. This is because some of the people from whom the hormone was prepared, must have been infected with CJD, even if they had shown no signs of the disease while they were alive. The use of growth hormone prepared from humans was banned in the UK in 1985.

3.2 People who have been treated with gonadotrophin hormones sourced from humans before 1973

Your medical records show that you were treated with gonadotrophin prepared from human pituitary glands before 1973. Many women were given this treatment, and four are known to have developed CJD worldwide.

This is because some of the people from whom the gonadotrophin was prepared, must have been infected with CJD, even if they had shown no signs of the disease while they were alive. The use of gonadotrophin prepared from humans was banned in the UK in 1973**.**

3.3 People who have been told by a specialist that they have a risk of developing an inherited (genetic) form of CJD or other human prion disease that runs in families

Someone in your family has an inherited (genetic) form of CJD or other human prion disease that runs in families. Inherited CJD is rare, and accounts for 15 out of every 100 cases of CJD in the UK.

A faulty gene causes inherited CJD disease, and this faulty gene can be inherited (passed) from parent to child. You should discuss your risk with a genetic specialist. If you are

carrying the faulty gene, then you could spread CJD to other patients if you donate blood, organs and tissue or have an operation. No cases have been reported of inherited CJD being spread to others in this way but it is possible that this form of CJD could infect other people.

Where can I find out more?

More information on CJD and what to do is contained in the leaflet 'Information for people who have an increased risk of CJD'.

The following organisations offer further information and support:

- CJD Support Network: www.cjdsupport.net. Helpline: 0800 0853527
- Public Health England: https://www.gov.uk/government/collections/creutzfeldt-jakob-disease-cjd-guidance-data-and-analysis
- Health Protection Scotland: http://www.hps.scot.nhs.uk/haiic/creutzfeldtjakobdisease.aspx
- National CJD Research and Surveillance Unit: www.cjd.ed.ac.uk
- The National Prion Clinic offers help and support to people with or at risk of CJD and their families: www.nationalprionclinic.org/
- The Institute of Child Health supports people who have received human derived pituitary growth hormone. Leah Davidson coordinates care for people affected by growth hormone related CJD. Tel: 020 7404 0536 Email: L.Davidson@ucl.ac.uk

We last updated this leaflet in June 2018. To check for any new information, please see the latest version at:
https://www.gov.uk/government/publications/cjd-information-leaflets-for-patients-and-healthcare-professionals
http://www.hps.scot.nhs.uk/haiic/creutzfeldtjakobdisease.aspx

We welcome feedback on this leaflet – please send your comments to:
cjd@phe.gov.uk or NSS.HPSInfectionControl@nhs.net

Published: July 2018
PHE publications gateway number: 2018233