CJD (Creutzfeldt-Jakob disease)

CJD - lyodura and the risk of exposure during healthcare - frequently asked questions

We would like to reassure all our patients that tight regulations govern all infection control processes at Addenbrooke's, and that we strictly adhere to all current guidelines including those to prevent CJD.

What is CJD?

Creutzfeldt-Jakob disease (CJD) is one of a rare group of diseases, known collectively as transmissible spongiform encephalopathies (TSEs). They affect the structure of the brain causing dementia and a range of neurological symptoms, and cause death.

A number of TSEs are recognised in both humans and 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: of these, sporadic CJD accounts for 85% of cases. The other types are familial ('inherited, genetic'), iatrogenic ('from healthcare') and variant CJD (vCJD).

1. Sporadic CJD is most common in those aged over 50 years, and affects about one person per million per year worldwide. It is thought to arise spontaneously. Familial CJD is an inherited genetic disease. The patients are often younger and the duration of the illness has a longer time course than sporadic CJD. Between six and ten cases are seen each year in the UK. The clinical features of
2. familial CJD are variable, even within affected families.
3. Iatrogenic CJD develops as the result of medical/surgical treatment, when the patient is exposed to body tissues from someone who has CJD (see below).
4. Variant CJD (vCJD) was first recognised in the UK in 1996. It is thought to be caused by dietary exposure to the BSE agent of cattle, although no one knows the exact route of infection. It is different from the other types of human CJD; it typically affects younger people, most often in their late 20s and its symptoms are different.

Is there a test for CJD in humans?

We are learning more about the clinical features of CJD, but we can only reliably diagnose
CJD by looking at the cells in the brain after a biopsy or as part of a postmortem. There is no simple test we can carry out on people who we think have CJD or those who might develop it in the future.

Is there a treatment for CJD?

There is no treatment for CJD currently available.

Who has developed variant CJD so far?

By September 2004, almost 150 cases of vCJD have occurred in the UK, and a small number in other countries. It is thought that the UK epidemic might have reached a peak but we don't know how many people will be diagnosed with this disease in the future.

How is the CJD acquired?

CJD is a very unusual disease because it is caused by an abnormally shaped protein (called prion). This 'bad' protein can make the body's normal prion proteins change shape, which causes the symptoms in the human. Once this has happened, further copies of the body's prion protein change shape - which looks like the spread of an infection. This happens very slowly, which accounts for the delay between encountering the 'bad protein' and developing the symptoms.

From a practical point of view, this can be considered as an 'infection' - although the protein doesn't have the same features as bacteria, viruses, funguses and other parasites that usually cause infections in humans.

This is a new disease, which is difficult to study. This means that experts find it difficult to give definitive advice. Most scientific evidence is gained from animal studies and epidemiology (the study of diseases in populations of humans). From animal studies (including the BSE epidemic in cattle), we know the 'bad' prion protein can enter the body and cause damage when it is eaten. We also know that it can cause problems when it is injected into the body. The amount of prion protein and how and when it is introduced seem to be very important, which helps us decide the risk of 'catching' CJD.

There is no evidence that CJD is spread between household members eg by coughing or sneezing, direct skin contact or sexual contact, nor is there evidence of transmission during pregnancy/birth from mother to baby.

Where did variant CJD come from originally?
The consumption of BSE-contaminated beef or other bovine-derived products remains the most likely means by which vCJD was acquired. This means that most of the UK population have been exposed to prion protein in their diet but it is currently anticipated that only a very few will ever suffer illness as a result.

**Which tissues of the body are most likely to cause CJD on exposure?**

Broadly, in vCJD, 'bad' prion proteins are found inside the nervous system and in the lymphoid (body's defence) tissues throughout the body. These are classified for risk according to the latest evidence - as of September 2004:

- 'High-risk' tissues in vCJD: central nervous system and posterior (back of) eye.
- 'Medium risk' tissues in vCJD the olfactory epithelium ('smell cells' at the top of the nose), anterior (front of) eye and cornea (covering of eye); gastrointestinal (gut) lymphoid tissue (including the tonsil, appendix and rectum).
- 'Tissues of concern' include the spleen, lymph nodes, thymus and adrenal gland.

**Have people acquired CJD during surgery?**

The brain and some other nervous tissue are most likely to have 'bad' prion protein in them, and are, therefore, potentially most likely to cause CJD.

All surgery carries risks and vCJD is a new risk. The risk comes from the transmission on surgical equipment from one CJD-infected patient to the next. The actual risk depends on the type of tissues coming into contact with the equipment and how easy it is to remove the 'bad' prion protein from the equipment. It is much more difficult to decontaminate medical equipment from 'bad' prion protein than to kill or remove bacteria and viruses.

This is one reason why hospitals (and dentists) have changed their infection control procedures since vCJD was first recognised. The type of decontamination methods have changed and, in some cases, disposable equipment is used.

**Have people developed vCJD after blood transfusions?**

By September 2004, there have been two cases of probable transmission of vCJD infection associated with blood transfusion in the UK (announced late in 2003 and 2004).

**How else have people contracted CJD due to healthcare?**

By September 2004, over 100 people have contracted CJD through treatment with
contaminated human growth hormone.

**What public health measures are being used to prevent CJD infection?**

Several public health measures have been implemented to minimise the risk of transmission of vCJD to humans from meat and meat products infected with BSE. These include banning the feeding of mammalian protein to other mammals, and removing certain high-risk tissues from the human food chain.

Other public health measures are aimed at minimising any possible risk of transmitting vCJD between people. These include:

- Measures to protect the blood supply,
- Improving decontamination standards for surgical instruments, and taking special infection control precautions when operating on patients who have, or are 'at-risk' of, vCJD.
- Although there is no evidence that vCJD can be sexually transmitted or transmitted from parent to child, as a precautionary measure, men who are 'at-risk of vCJD for public health purposes' are advised not to be sperm donors.

**Can I acquire CJD in hospital?**

There is a very low risk of acquiring CJD as a result of hospital and/or medical treatment and actual cases are very rare. This is because you need to come into the right type of contact with enough of the 'bad' prion protein from one of the rare people who have got or are developing CJD.

Also, hospital and public health experts are being very cautious and taking all known measures to reduce the risks within healthcare.

The reported healthcare-related cases of CJD are most likely to have occurred through exposure to contaminated instruments, through transplantation, blood transfusion or human-derived growth hormone or potentially some human-derived plasma products.

**Can I get CJD from GIVING blood products including transfusions?**

You can't catch CJD from giving blood or blood products - all the equipment used is disposable.

**Can I get CJD from RECEIVING blood products including transfusions?**
The risk of catching CJD by receiving blood or blood products remains uncertain. Each year, we learn more about this disease, how to prevent it and how to advise people who might have been exposed to it.

As of September 2004, there have been no recorded instances of vCJD being spread through plasma products from individuals who later developed vCJD.

From time to time, certain groups are people are contacted by their doctors to warn them that they might be ‘at risk’ and what they can do next. This is because doctors and other experts are being very cautious.

Up to September 2004, nine people who have donated plasma in the UK are known to have developed vCJD - in total they have made 23 plasma donations. The donated plasma has been used to manufacture some batches of some blood products in some UK companies. Some of these products might carry a risk of CJD, depending on how, when and where they were prepared.

Not all plasma (or blood-derived) products are thought to be risky - for many, the amount of potential 'infectiousness' is so low that patients receiving them do not need to take any special precautions. These 'no-risk' products include the 'anti-D' used in Rhesus negative pregnant women, and intramuscular immunoglobulin used in travel vaccinations against hepatitis A.

**What measures are being taken to prevent CJD infection through blood products?**

Because of the (low) risk of catching CJD from blood and blood products, several public health precautions have been taken to reduce any possible risk of transmitting CJD through blood. As of September 2004, these precautionary measures include:

- Withdrawal and recall of any blood components, plasma products or tissues obtained from any individual who later develops vCJD.
- Importing plasma from the USA for making plasma products.
- Removal of white blood cells (they carry the greatest risk of transmitting vCJD) from all blood used for transfusion.
- Importing fresh frozen plasma from the United States for use in patients born on or after 1 January 1996.
- Not accepting donations of blood or plasma from people who have themselves received a blood transfusion in the UK since 1980 (or from those who are not sure).
Promotion of appropriate use of blood and tissues and alternatives throughout the NHS.

How do hospitals reduce risks to patients?

Hospitals follow guidance given by national expert bodies on CJD. This includes guidance on decontamination measures for medical/surgical equipment. Much effort has gone into ensuring that the decontamination of all surgical instruments is to the highest standards.

They take additional measures when they are treating people who are suspected to have CJD or are in 'at risk' groups - for example they can use disposable equipment or destroy it after use on an 'at risk' patient.

How do hospitals contact people who they think might be at an increased risk?

A national expert group assesses the risk to various groups of patients (eg people who have received blood products or have had types of surgery), and advises whether they should be contacted and informed about their possible exposure. This depends on the level of risk for that group. Their own doctors contact them by post and invite them to discuss their options with a trained member of staff.

One reason for contacting patients is so they can take measures to prevent any additional 'bad' prion protein getting into the healthcare system.

What does being 'at risk' mean?

People usually want to know from their doctors 'for certain' whether they will or will not develop a condition. Unfortunately, this is not always possible.

vCJD is a very difficult disease to predict risk because it is new and difficult to understand. Most people in the UK will have been exposed to the 'bad' prion protein through their diet - but only some people have an additional risk because of their exposure through healthcare treatments. We can not yet estimate the risk to an individual from diet but we can identify some of those at risk from healthcare procedures by careful examination of their medical records.

No doctor wants to worry patients unnecessarily but it is thought to be important to tell patients that they might be at an additional risk so that they can help their doctors to protect others from acquiring vCJD via the healthcare system.
What affect will being 'at risk' have on that patient's care?

The clinical care of the patients identified as 'at-risk' for public health purposes should not be compromised in any way. They might just have additional infection control measures during any surgery.

**What does an 'at risk' person need to do (or not do)?**

Prion protein (PrP) is a naturally occurring part of the brain and other parts of the body in both humans and other animals. It is an abnormal form of this prion protein that is thought to cause CJD, and becomes 'infectious' to other proteins and animals.

We don't yet understand what this prion protein does, but when it is abnormal the protein is resistant to normal breakdown in the cell. The abnormal protein clogs up the cells, killing some of them (especially in the central nervous system, including the brain), which causes sponge-like ('spongiform') holes in the tissues.

One 'changed' prion protein molecule seems to act as a template for change in further molecules of the prion protein. The first abnormal protein molecule might have arisen 'by chance' (ie in sporadic CJD), due to a mutation in the genetic code for the protein (ie in familial CJD) or because of exposure to external prion proteins by diet or in healthcare (ie in vCJD).

**What is a prion protein and what makes it abnormal in CJD?**

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Where can I find out more about CJD?

vCJD is a new disease and the information and advice about it changes often.

Useful Links:

CJD Support Network
for people affected by CJD

National CJD Surveillance Unit, Edinburgh
lots of useful information, especially for researchers

Department for Environment, Food and Rural Affairs
all about BSE

CJD - Lyodura in neurosurgery

What is lyodura and why was it used in neurosurgery?

Lyodura is processed dura mater (dura being the tough outside membrane that covers the brain), which was used between 1981 and 1992 to seal wounds following neurosurgery.

Lyodura and CJD

The use of lyodura has been associated with the transmission of the infectious agent of Creutzfeldt-Jakob disease (CJD); however, the risk of CJD infection as a result of the use of lyodura remains extremely low.

When was lyodura used at Addenbrooke's?

Lyodura was used in neurosurgery at Addenbrooke's between 1981 and 1992. During this time, lyodura was commonly used by surgeons worldwide and no adverse side effects of lyodura were known.

Lyodura is no longer used in neurosurgery at Addenbrooke's. Patients who have had neurosurgery at Addenbrooke's since 1992 are not at risk of CJD from this medical reagent.

I had neurosurgery between 1981 and 1992, who can I contact for advice?
We have set up a helpline for any patients (or their relatives) who have undergone neurosurgery at Addenbrooke's between 1981 and 1992 (when lyodura was in use).

If they have any concerns at all, they can contact this free phone number. They will be able to talk in confidence about their case and receive further information.

Helpline: 0800 389 8625 (open Monday to Friday 0900-1800 hrs)