Patients at increased risk of Creutzfeldt-Jakob Disease

Actions for healthcare staff

This leaflet is for healthcare staff caring for patients who are at increased risk of Creutzfeldt-Jakob Disease (CJD). There is also a 'Background information for healthcare staff' leaflet and two patient information leaflets 'Information for people who have an increased risk of CJD' and 'Who has an increased risk of CJD?'.

1 The CJD Incidents Panel and Incidents

1.1 The CJD Incidents Panel

The CJD Incidents Panel advises on how to manage the possible risk of CJD transmission between patients. The CJD Incidents Panel may advise that instruments used on a patient with or at risk of CJD should be quarantined immediately, be destroyed or sent for research. The Panel also advises whether or not patients should be contacted and informed about their possible exposure.

The CJD Section at the HPA Centre for Infections provides the secretariat for the Panel, and provides help to healthcare teams implementing Panel advice.

1.2 CJD healthcare incidents

CJD 'incidents' occur when there is a possibility that patients could have been exposed to CJD through:

- exposure to contaminated surgical instruments
- organ or tissue transplants
- blood transfusions
- treatment with plasma products such as clotting factors or albumin.

The CJD Incidents Panel advises how to manage these incidents, and how to manage patients who could have been exposed to CJD. Local infection control teams and health protection teams should seek advice from the CJD Incidents Panel on how to manage these incidents.

More information on the CJD Incidents Panel is available on the HPA website. This includes the CJD Incidents Panel framework document, which sets out the principles of managing CJD incidents and also describes the risk assessment models that underpin the risk management of surgical and blood incidents.

1.3 The Panel's recommendations

The Panel advises on the need to contact patients who could have a risk of at least 1% of being infected with CJD through medical procedures. These patients should be told they are at increased risk of CJD and that they should take certain safety measures as a precaution to prevent CJD being spread to other patients.

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

1.4 Health protection teams

Patients who are at increased risk of CJD may have had surgery since the time when they were exposed to CJD. Any instruments that have been used on tissues with medium-low, medium-high or high infectivity, could still transmit CJD to other patients (Table 1). This is because infectious prion proteins on the surgical instruments are not completely removed by the usual decontamination processes.

The estimated risk of transmitting CJD from one patient to another via surgical instruments decreases each time the instruments are used and decontaminated. After being used and decontaminated around 10 times (20 times for high risk tissue), the risk from those instruments is probably reduced.

Health protection teams should:

- Check whether patients have had surgery involving medium-low, medium-high, and high risk tissues during the last 12 months. This includes patients who are at increased risk who have died within the previous year.
- Health protection teams should report any surgery carried out on medium-low, medium-high, and high risk tissues involving instruments that may have been used and decontaminated fewer than 10 times (20 times for high risk tissues) since being used on patients at risk of CJD.
- Check whether patients have donated blood, organs or tissue since they were exposed to an increased risk of CJD.
- Complete a CJD Incidents Panel surgical incident reporting form if appropriate.

Surgical departments re-use instruments at different rates. Reviewing the medical history over the last 12 months for each patient who is at increased risk of CJD should reveal any instruments that could still transmit CJD to other patients.

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

Table 2 Tissue infectivity levels for patients with, or at increased risk of, CJD

Infectivity level	Tissues of patients with, or at risk of CJD
High	Brain; spinal cord; dura mater; cranial nerves, specifically the entire optic nerve and only the intracranial components of the other cranial nerves; cranial nerve ganglia; posterior eye; pituitary gland
Medium-high	Olfactory epithelium Tonsil, spleen (only in variant CJD)
Medium–low	Anterior eye Fixed lymphoid tissue (only in variant CJD)

Once an incident has been reported to the CJD Incidents Panel, the health protection team should continue to work with the CJD Incidents Panel and local healthcare professionals to ensure that appropriate public health investigations and actions are carried out.

1.5 Infection control teams

If a patient at increased risk of CJD is due to undergo an invasive medical or surgical procedure, including endoscopy, infection control teams should follow guidance prepared by the ACDP TSE Working Group, 'Transmissible spongiform encephalopathy agents: safe working and the prevention of infection'.

This describes how to reduce the risk of CJD infection in hospitals when carrying out certain surgical and investigative procedures, and in community healthcare settings. Annex E of the guidance relates to the quarantining of instruments, and Annex F relates to endoscopy procedures.

1.6 Clinicians informing patients that they are at increased risk of CJD

The GP is usually best placed to inform patients that they are at increased risk of CJD. In some cases a specialist doctor who provides ongoing care may inform a patient. In these cases, the specialist should also inform the GP of the patient's increased risk status and that public health actions are required.

When discussing CJD risks with a patient, it is important to communicate two messages. First, that the risk of the patient being infected with CJD is uncertain, but is likely to be low. Second, that it is important that the patient should follow advice to reduce any risk of the infection spreading to other patients.

Two patient leaflets 'Information for people who have an increased risk of CJD' and 'Who has an increased risk of CJD?' should be given to patients during these consultations.

Patients may find the news that they are at increased risk of CJD both distressing and difficult to understand. They may want an absolute guarantee that they will not develop CJD. This is clearly not possible.

Many patients are likely to need more than one opportunity to discuss what this means for them if they are to come to terms with what they have been told. It may be helpful to consult a trained counsellor for advice on managing this process.

The healthcare professional informing a patient of their increased CJD risk status may wish to arrange follow up visits to give the patient opportunities to discuss these complex issues with appropriate staff.

1.7 General Practitioners

The patient's GP should:

- Record in the patient's primary care records that the patient is at increased risk of CJD, the reason for this, and that special infection control measures may be needed for medical and surgical procedures, including endoscopy.
- Include this information in any referral letters if the patient needs surgery, including specialist dental surgery, or other invasive procedure.
- Pass this information to any specialist doctors providing ongoing care to the patient.
- Check if the patient has had surgery in the past. If this is the case, the GP should tell the local Health Protection Team, who will take action if necessary.

1.8 Surgical and other hospital staff

Healthcare staff should ensure that patients who are at increased risk of CJD are not subjected to any delays or postponement of treatment as a result of uncertainty regarding the public health actions required.

1.9 Dentists

Patients who are at increased risk of CJD require the same standards of infection control as any other patients.

The only difference in the care of a patient at increased risk of CJD is that dentists should include their increased CJD risk status in referral letters to specialists such as maxillofacial surgeons. This is why patients at increased risk of CJD are asked to inform their dentist.

Dentists should be aware of the guidance prepared by the ACDP TSE Working Group, 'Transmissible spongiform encephalopathy agents: safe working and the prevention of infection'. This guidance 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.

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 who are at increased risk of CJD[1]. The CJD Incidents Panel and HPA have prepared an advice note on the dental care of individuals who are at increased risk of CJD[2].

[1] http://www.dh.gov.uk/en/Publicationsandstatistics/Lettersandcirculars/Dearcolleagueletters/DH_4102752

[2] www.hpa.org.uk/web/HPAwebFile/HPAweb_C/1211788871050

2. People who are at increased risk of CJD (see Figure 1)

2.1 Patient groups

Several groups of people are at increased risk of CJD. Everyone in these groups should follow advice to reduce the risk of the infection spreading to other patients.

Table 1 People at increased risk of CJD

The following groups of people are at increased risk of CJD:
Related to blood transfusions
People who have received blood from someone 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 someone who has also given blood to a patient who went on to develop variant CJD
Related to surgery
People who have had surgery using instruments that had been used on someone who developed 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
Related to other medical care
People who have been treated with UK sourced plasma products between 1980 and 2001
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 the genetic form of CJD

2.2 Actions for people at increased risk of CJD

All people who are at increased risk of CJD are asked to help prevent any further possible transmission to other patients by following this advice:

- 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, dental or surgical procedures, tell whoever is treating you beforehand so they can make special arrangements for the instruments used to treat you if you need certain types of surgery or investigation.
- You are advised to tell your family about your increased risk. Your family can tell the people who are treating you about your increased risk of CJD if you need medical or surgical procedures in the future and you are unable to tell them yourself.

Medical care abroad for people at increased risk of CJD

If a patient at increased risk of CJD needs an operation or endoscopy while abroad, they should:

- Tell the medical staff that, in the UK, doctors may take special infection control measures during surgery and endoscopy to reduce the risk of infecting other patients with CJD. Guidance on these infection control precautions is available on the HPA website www.hpa.org.uk/CJD
- Tell the medical staff to 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 plan to do, or have already done.

The HPA duty doctors are available 24 hours a day every day. They 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 undertake the procedure, the patient should tell them as soon as possible afterwards. This may happen in the UK as well as abroad, for example, if the patient was unconscious and needed immediate treatment. Procedures for controlling infection can still be effective, even after the medical procedure.

3 Types of incidents

3.1 Surgical CJD incidents

Surgical (or other) instruments may be contaminated with prion protein when they come into contact with 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 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[3]. The risk assessments show that the risk to patients is highest when surgical instruments have only been used and decontaminated a small number of times after being used on a patient with, or at increased risk of, CJD.

The Panel's advice on whether to contact patients following a CJD 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, or at increased risk of, CJD
- the infectivity levels of the tissues on which the instruments have been used
- the type of instruments used
- the processes in place for decontaminating the surgical instruments
- whether the instruments can be traced.

The Panel currently advises that, in general, there is no need to contact patients who have been exposed to instruments or blood, tissues or organs if the index patient is at increased risk of CJD. There are exceptions to this e.g. if the index patient has received variant CJD implicated blood components, or is at risk of genetic CJD.

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[3] Assessing the risk of vCJD transmission via surgery: an interim review, Department of Health, 15th June 2005

3.2 Blood related CJD incidents

3.2.1. Blood components

Four people have been infected with variant CJD following a blood transfusion. Three of these people developed symptoms of variant CJD and died from clinical variant CJD. All four cases had received transfusions of non-leucodepleted red blood cells between 1996 and 1999.

Patients who receive blood transfusions from donors who later developed variant CJD may have a risk of variant CJD infection. Certain public health precautions are recommended for these recipients, and they are traced and notified that they have an increased risk of variant CJD.

Some variant CJD cases are found to have received blood donations in the past (Figure 1). If the people who donated blood to these cases were incubating variant CJD at the time of donation, then they could have been the source of the recipients' variant CJD infection. This possibility has been investigated in a risk assessment carried out by the Department of Health's Standards and Quality Analytical Team[4] . In 2005 the CJD Incidents Panel considered this risk assessment, and recommended that these blood donors should be informed of their risk of variant CJD infection and considered to be at increased risk of variant CJD.

Some of these blood donors have given blood to other patients (Figure 1). In September 2005, the CJD Incidents Panel considered the risk to patients who had received blood from donors to variant CJD cases. The risk of variant CJD infection in this group is very uncertain. However, some of these recipients may have a risk of variant CJD infection and could pose a risk to others. The CJD Incidents Panel has recommended such recipients have an increased risk of variant CJD, if the probability of being infected with variant CJD is estimated to be greater than 1%.

Neither the threshold, nor the exact risk estimate value for any individual recipient should be used as an indicator of an individual's risk of developing variant CJD. This risk is unknown.

3.2.2 Patients with bleeding disorders

A person with haemophilia has been found to have evidence of infection with the vCJD abnormal prion protein only in his spleen at post mortem. The post mortem was carried out as part of a study jointly co-ordinated by the UK Haemophilia Centre Doctors Organisation and the National CJD Surveillance Unit.

This haemophilia patient had been treated in the 1990s with several batches of UK-sourced clotting factors, including one batch of factor VIII that was manufactured using plasma from a donor who went on to develop vCJD. The plasma donor developed symptoms of vCJD 6 months after donating the plasma in 1996. The haemophilia patient was in his 70s when he died of a condition unrelated to vCJD, 11 years and one month after receiving the batch of implicated

[4] Assessing the implications for blood donors if recipients are infected with vCJD. Department of Health. July 2005.

factor VIII. He had no signs or symptoms of vCJD or other neurological disease when alive.

A final view as to how vCJD abnormal prion protein was transmitted to this haemophilia patient has yet to be reached because investigations are continuing to be sure of the source.

This is the first time that vCJD abnormal prion protein has been found in a patient with haemophilia, or any patient treated with plasma products

This finding does not change existing public health vCJD 'at risk' status for patients with bleeding disorders.

Patients who had received plasma products manufactured from plasma donated by individuals who later developed variant CJD were notified of this in 2004 and 2006. Plasma from these donors had been used to manufacture factor VIII, factor IX, antithrombin, intravenous immunoglobulin G, albumin, intramuscular human normal immunoglobulin and anti-D.

Patients treated with these plasma products are managed according to an assessment of potential variant CJD infectivity carried out by the Health Protection Agency with the Panel. Plasma product infectivity data are combined with batch manufacturing data to calculate the likely infectivity of each batch of the implicated plasma products.

Three groups of patients are involved: patients with bleeding disorders, patients with primary immunodeficiency disease, and patients with other disorders who had been treated with sufficient quantities of plasma products to result in a greater than 1% risk of variant CJD infection.

The UK Haemophilia Doctors and patient representatives, the Panel and UK Health Departments agreed that it was likely that many patients with bleeding disorders would have had a sufficient exposure to these implicated plasma products to put them'at increased risk of variant CJD. It was also thought likely that further batches of UK-sourced plasma products would be implicated in the future as more cases of variant CJD arose.

Therefore all patients with bleeding disorders[5] who were treated with UK-sourced pooled factor concentrates or antithrombin[6] between 1980 and 2001[7] were informed that they were at increased risk of variant CJD and that special precautions should be taken. This included many patients who had not received plasma products sourced from known variant CJD patients.

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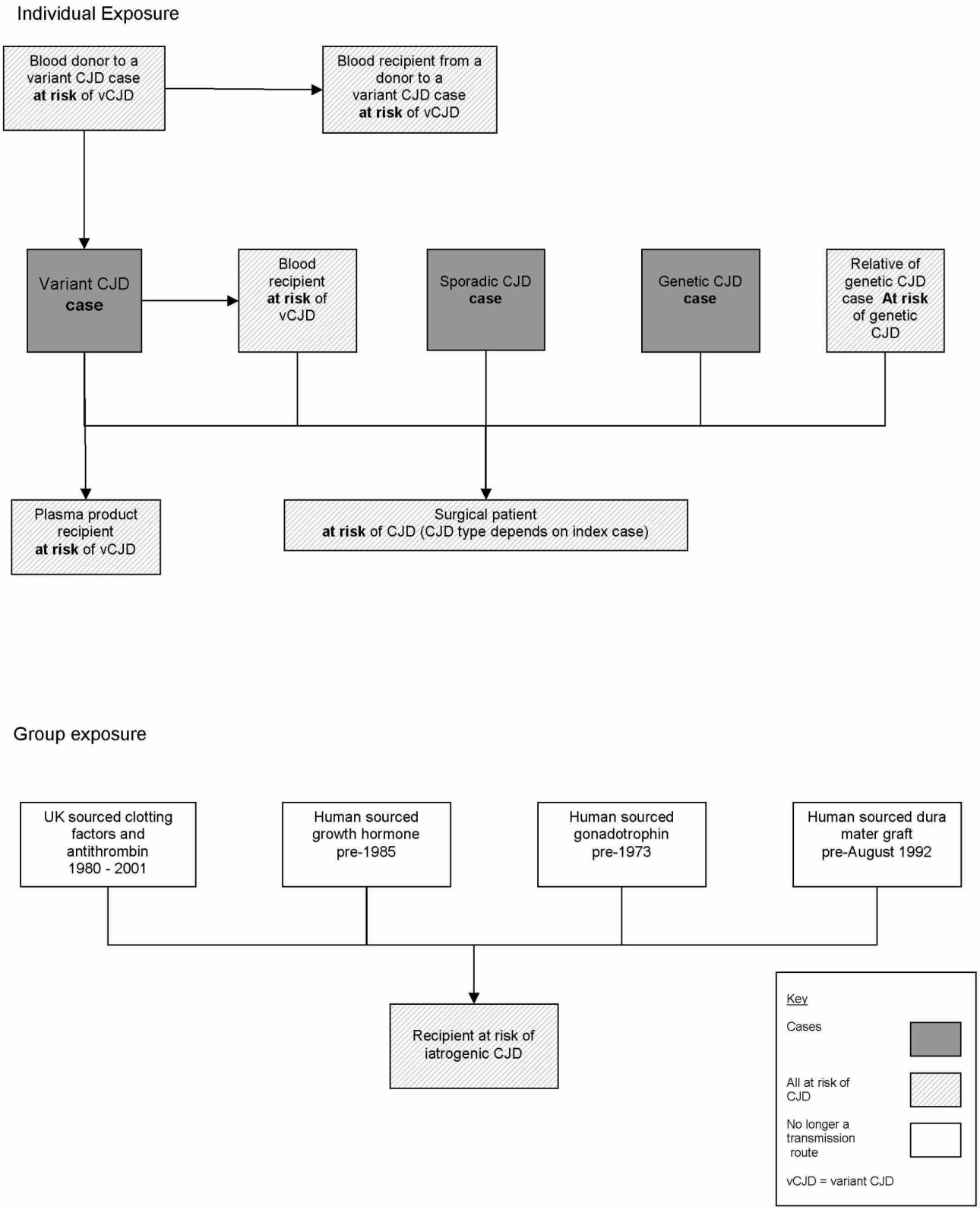
[5] Defined here as congenital and acquired haemophilia (Haemophilia A and Haemophilia B), Von Willebrand Disease, other congenital bleeding disorders and congenital antithrombin III deficiency.

[6] ie. clotting factors and antithrombin made from pooled plasma. These include factor VIII, factor IX, factor VII, factor XI, factor XIII and prothrombin complex concentrates as well as antithrombin.

[7] The start date of 1980 is when BSE is thought to have entered the human food chain. The end date of 2001 is the last possible expiry date of any product manufactured by the UK fractionators that was sourced from UK donors until 1998.

Patients who have been treated with other types of plasma products were assessed on an individual basis, to see if they had an increased risk of developing variant CJD.

Figure 1: Relationships between patient groups at risk of CJD



4 Useful websites

You can get more information about CJD from the following websites:

CJD Incidents Panel
http://www.hpa.org.uk/CJDIncidentsPanel

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

Department of Health information for dentists
http:www.dh.gov.uk/PublicationsAndStatistics/LettersAndCirculars/DearColleagueLetters/DearColleagueLettersArticle/fs/en?CONTENT_ID=4102752&chk=7HspA

National CJD Surveillance Unit
http://www.cjd.ed.ac.uk/index.htm
Phone: 0131 537 2128

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

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

Institute of Child Health: 30 Guilford Street, London WC1N 1EH
Leah Davidson coordinates care for people affected by growth hormone related iatrogenic CJD
Tel: GRO-C Email: L.Davidson@ GRO-C

CJD Therapy Advisory Group guidance:
http://www.dh.gov.uk/en/Policyandguidance/Healthandsocialcaretopics/CJD/CJDgeneralinformation/DH_4031039

Department of Health
http://www.dh.gov.uk/en/Policyandguidance/Healthandsocialcaretopics/CJD/CJDgeneralinformation/index.htm

Spongiform Encephalopathy Advisory Committee
www.seac.gov.uk/
Department for Environment, Food and Rural Affairs BSE home page
http://www.defra.gov.uk/animalh/bse/index.html

We last revised this leaflet in February 2009. To check for any updates to this information, please see the current version of this leaflet at www.hpa.org.uk/CJD
We welcome your comments on this leaflet. Please send them to cjd@hpa.org.uk