Policy Statement
To ensure all relevant clinical staff are aware of the risks associated with prion disease and take appropriate steps to prevent the risk of transmission when clinical interventions are to be undertaken.

This policy applies to all staff employed by NHS Greater Glasgow & Clyde and locum staff on fixed term contracts.

KEY CHANGES FROM THE PREVIOUS VERSION OF THIS POLICY

- Removal of multiple transfusion question
- Change in precautions for use of endoscopes on those with vCJD

Document Control Summary

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<td>Infection Prevention and Control Policy Sub-Group</td>
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www.nhsggc.org.uk/infectionpreventionandcontrol
1. Responsibilities

All staff must:
- Follow this policy.
- Inform a member of the Infection Prevention Control Team (IPCT) if this policy cannot be followed.
- Report to line managers any deficits in relation to knowledge of Transmissible Spongiform Encephalopathy (TSEs), facilities/ equipment or incidents that may have resulted in cross-contamination.

Managers must:
- Ensure if appropriate that all staff have education/ training on the principles of managing patients with TSEs.
- Ensure that adequate resources are in place to allow for the recommended infection control measures such as Personal Protective Equipment (PPE) to be implemented.
- Support staff in any corrective action or interventions if an incident occurs that may have resulted in cross-contamination.

The Infection Prevention Control Team must:
- Provide education opportunities for staff on the contents of this policy.
- Act as a resource for guidance and support when advice on TSE/ Creutzfeldt-Jakob Disease (CJD) is required.
- Provide advice on individual risk assessments for TSE/ CJD decisions.
- Keep this policy up-to-date.
2. Introduction

TSEs are rare neurodegenerative conditions that include:
- Gerstmann-Straussler-Scheinker syndrome,
- Fatal familial insomnia
- Creutzfeldt–Jakob Disease.

CJD is the abbreviation used throughout this document as the other TSEs are very rare and the best practice principles will apply equally to these conditions. The agents responsible for these diseases are proteins, which become altered in their configuration and are known as “Prions”. CJD may be passed on in families (10-15% of all cases) or a spontaneous mutation may occur giving rise to sporadic cases with no associated risk factors.

A variant form of CJD was identified in 1996 which was described as being identical to Bovine Spongiform Encephalopathy (BSE or “Mad Cow Disease”). It was thought to have been transmitted to humans by eating contaminated meat products. Other instances of transmission of CJD have been described via receiving blood products and occasionally through instruments contaminated with CJD. Experts have considered all these factors and guidance is constantly reviewed and updated.

Unlike micro-organisms, prions are not destroyed by conventional methods of decontamination and sterilisation, therefore a risk assessment has to be undertaken before any clinical procedure where there is considered to be a risk of potential transmission. (See Appendix 1 Algorithm)

The guiding principle in all cases is the attention to:

“best infection control practice” and well informed healthcare workers (HCWs) as to the risk.

The current UK CJD guidelines are regularly reviewed and updated especially as more evidence becomes available.

3. General Information

3.1. Identified forms of Creutzfeldt-Jakob Disease (CJD)

Classical CJD

Familial
found in first-degree relatives

Sporadic
occurs in 1:1,000,000 of the population with no apparent risk factors

The most up-to-date version of this policy can be viewed at the following website:

www.nhsggc.org.uk/infectionpreventionandcontrol
Iatrogenic CJD
Worldwide, cases of iatrogenic CJD have been associated with the administration of hormones prepared from human pituitary glands and dura mater preparation and one definite case has been reported associated with a corneal graft (it is possible that the corneal tissue was contaminated by posterior segment tissue during processing). Iatrogenic transmission has also been identified following neurosurgical procedures with inadequately decontaminated instruments or EEG needles.

Variant CJD (vCJD)
Found to be pathologically identical to BSE. Clinical presentation differs from Classical forms of CJD. There is evidence that vCJD can be transmitted via exposure to contaminated blood/blood products (since 2003, four cases (3 clinical and 1 asymptomatic) of presumed person to person transmission of vCJD infection via blood transfusion of non-leuco-depleted red blood cells have been reported in the UK. (In 2009 a case of probable asymptomatic vCJD infection via plasma products was reported in a haemophiliac). There is a theoretical risk of transmission of vCJD from contaminated surgical instruments.

3.2. Diagnostic Criteria for CJD and vCJD (see Appendix 3)

3.3. Categorisation of cases and at risk groups (see Appendix 4)

Cases are symptomatic patients who fulfil the diagnostic criteria for definite, probable or possible sporadic, familial or acquired CJD as described above. Patients in risk groups do not have symptoms of CJD, rather they have been characterised as at risk on the basis of a family history or genetic testing, or a previous exposure.

3.4. Reportable disease

The patient’s clinician will report any patient suspected on clinical grounds of having vCJD/ CJD to the Clinical Team at the National Surveillance Unit in Edinburgh Tel: 0131 332 2117. Patients identified as at risk of CJD (see below) should be reported to the Public Health Protection Unit (PHPU) 0141 201 (6)4917 e-mail PHPU@ggc.scot.nhs.uk who will report them to HPS.

3.5. At risk groups

Patients identified to be at increased risk include those with risks in relation to:
The Management of Patients with Transmissible Spongiform Encephalopathy (TSE) including all forms of Creutzfeldt-Jakob Disease (CJD)

Effective From April 2014
Review Date April 2017
Version 5

The most up-to-date version of this policy can be viewed at the following website: www.nhsggc.org.uk/infectionpreventionandcontrol

Transfusion of blood/ blood components
- Individuals who have been identified as having received blood or blood components from 300 or more donors since January 1990.
- Individuals who have received labile blood components (whole blood, red cells, white cells or platelets) from a donor who went on to develop vCJD.
- People who have given blood or blood components to someone who went on to develop vCJD.
- People who have received blood or blood components from someone who has also given blood or blood components to a patient who went on to develop vCJD.

Surgery on high or medium risk tissue
- People who have had surgery using instruments that had been used on someone who developed CJD.
- People who have had an intradural neurosurgical or intradural spinal procedure before August 1992 who received (or might have received) a graft of human derived dura mater are at increased risk of transmission of sporadic CJD (unless evidence can be provided that human derived dura mater was not used).
- People who have received an organ or tissue from a donor infected with CJD or at increased risk of CJD.

Other medical care
- People who have been treated with certain UK sourced plasma products between 1980 and 2001.
- People who have been treated with growth hormone sourced from humans (before 1985). Human derived growth hormone may have continued in other countries after this date.
- People who have been treated with gonadotrophin sourced from humans (before 1973). Human derived gonadotrophin may have continued in other countries after this date.
- People who have been told by a specialist that they have a risk of developing the genetic form of CJD.

Patients at increased risk from genetic forms of CJD
- Individuals who have been shown by specific genetic testing to be at significant risk of developing CJD.
- Individuals who have a blood relative known to have a genetic mutation indicative of genetic CJD.
- Individuals who have two or more blood relatives affected by CJD or other prion disease.

The most up-to-date version of this policy can be viewed at the following website: www.nhsggc.org.uk/infectionpreventionandcontrol
3.6. **Questions to be asked of all patients undergoing Surgery/ Endoscopy**

All patients to undergo an invasive procedure will be asked the single question, i.e. they will be asked if they have been notified that they are at risk of CJD for public health purposes. All patients must be asked when being consented for any kind of surgery/ endoscopy. Subsequent actions depend on the patient’s response and the risk status of the tissues involved.

**Procedures should not be delayed whilst information is being collected and clinicians should be careful not to prejudice overall patient care.**

3.7. **Questions to be asked of all patients undergoing invasive procedures involving contact with high-risk tissues**

All patients undergoing invasive procedures on high-risk tissues must be asked the single question and three supplementary questions:

1. Have you been notified that you are at risk of CJD for public health purposes?
2. Have you a history of CJD or other prion disease in your family? If yes, please specify.
3. Have you ever received growth hormone or gonadotrophin treatment? If yes, please specify: i) whether the hormone was derived from human pituitary glands, ii) the year of treatment, and iii) whether the treatment was received in the UK or in another country.

4. **Transmission Risk**

Direct person-to-person spread of CJD and vCJD has not been shown to be a risk. Epidemiological evidence suggests that standard infection control precautions should be undertaken for all caring needs of CJD cases or those deemed at risk for CJD/ vCJD.

4.1. **Precautions to reduce the risk from blood/blood products/other human tissue derived products**

In the UK there are now measures in place to reduce this risk:

- Blood components, plasma products or tissues obtained from an individual who later develops vCJD are withdrawn to prevent their use.
- Plasma for the manufacture of plasma products, such as clotting factors, has been obtained from non UK sources since 1998.

The most up-to-date version of this policy can be viewed at the following website: [www.nhsggc.org.uk/infectionpreventionandcontrol](http://www.nhsggc.org.uk/infectionpreventionandcontrol)
4.2. **Transmission based precautions for cases of CJD and those identified as at risk**

Ward procedures and normal routine clinical contact does not pose a risk to HCWs, relatives or others in the community. These patients may be nursed in the open ward areas or in community settings. Additional precautions would only be required if specific interventions were to be undertaken involving contact with high or medium risk tissues (see appendix 2).

4.2.1. **Personal Protective Equipment (PPE)**

For any procedure involving the likelihood of aerosols or splashing, the following should be worn:

- disposable aprons/ waterproof gowns
- eye protection/ and masks or visors
- gloves

These should be disposed of as clinical waste at the end of the procedure.

4.2.2. **Body fluids**

Current evidence suggests there is no additional risk in saliva, excreta and other body secretions and these should be treated with the standard infection control precautions. However it is recommended that only trained staff that are aware of the hazards should carry out invasive procedures that might lead to contact with infective tissues.

4.2.3. **Spillages**

Standard infection control precautions for body fluid spillages should be used. Treat cerebrospinal fluid spillages the same as a blood spillage.

The most up-to-date version of this policy can be viewed at the following website:

[www.nhsggc.org.uk/infectionpreventionandcontrol](http://www.nhsggc.org.uk/infectionpreventionandcontrol)
5. Clinical Interventions

5.1. Precautions to reduce the risk from contaminated re-usable surgical instruments

Since prions cannot be destroyed by current decontamination procedures, special precautions must be taken for re-usable surgical instruments which come into contact with high infectivity tissues (neurological including spinal cord, and posterior eye) or medium risk tissue (including spinal ganglia, lymphoid reticular tissue and olfactory epithelium) of a patient who has been categorised as at risk (via either the single question or supplementary questions) (Appendix 2).

- For an invasive procedure on a person identified as at risk via the single question, for procedures involving contact with medium risk tissue, further information should be sought in the first instance. Infection Control should be contacted, if available and there is time, to conduct a full risk assessment. Generally the risk assessment takes account of the type of CJD/ vCJD of which the patient is at risk (Appendix 4); the types of tissue involved and, in some cases, whether the procedure is invasive. The result of the risk assessment will determine whether special precautions should be used. If a risk is confirmed the instruments used should be either i) single use, ii) quarantined after use if re-usable (and destroyed if the risk is confirmed) or, rarely, iii) kept for exclusive use on the same patient (most commonly endoscopes). If the patient answers yes and Infection Control are not available, or time is of the essence, adopt a precautionary approach and quarantine all re-usable instruments until the full risk assessment can be undertaken. If the patient is unable to respond, the procedure should go ahead using normal infection prevention control measures unless other risk information is available, e.g. GP letter (and unless it involves contact with high-risk tissue). If a risk is identified from the, e.g. GP letter or the procedure involves contact with high-risk tissues the re-usable instruments should be quarantined and if the risk is confirmed, dealt with as detailed below.

When a patient is identified as at risk or the risk remains unknown via the single question and/or supplementary questions and the invasive procedure involves contact with high-risk tissues, the instruments used should be either i) single use, or ii) quarantined after use if re-usable (and destroyed if the risk is confirmed) or, rarely, iii) kept for exclusive use on the same patient. For further details see Annex J at:

For those patients where the risk is confirmed:

- Contact Infection Control who will in turn contact PHPU. PHPU will inform HPS for surveillance purposes.
- Infection Control will discuss the risk with the patient (using patient information leaflet) and give the patient the usual infection control and public health advice. This advice is for people who have been identified as being at increased risk of CJD to reduce the risk of spreading CJD to other people:
  - Do not 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.
  - Do not 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.
  - You are advised to 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 patient’s GP should be informed and advised to record the patient’s CJD risk status in their primary care records. The GP should also include this information in any referral letter should the patient require invasive surgical, medical or dental procedures.
- Infection Control will inform medical records so that the electronic records can be flagged.
- Infection Control and Public Health will work together to identify any previous procedures which require to be reported to HPS.

5.2. **NICE Guidance**

The National Institute for Clinical Excellence (NICE) has recommended a number of steps to prevent transmission of CJD/ vCJD from those who were potentially exposed from contaminated beef to those who were not. [http://www.nice.org.uk/nicemedia/live/11332/31756/31756.pdf](http://www.nice.org.uk/nicemedia/live/11332/31756/31756.pdf)

NICE guidance covers management of all patients undergoing procedures involving instruments and endoscopes that might pose a risk of transmission of CJD/vCJD. It does not cover dental procedures. The guidance includes recommendations for invasive procedures involving contact with high-risk tissues:

The most up-to-date version of this policy can be viewed at the following website: [www.nhsggc.org.uk/infectionpreventionandcontrol](http://www.nhsggc.org.uk/infectionpreventionandcontrol)
• Instruments that come into contact with high-risk tissues must not move from one set to another.

• Supplementary instruments which come into contact with high-risk tissues should be either: i) single use or ii) should remain with the set to which they have been introduced.

• Rigid neuroendoscopes should be used where possible. They should be able to be autoclaved after use.

• All neuroendoscope accessories should be single use.

• A separate pool of neuroendoscopes and re-usable surgical instruments should be used on children born since 1 January 1997 and who have not previously undergone procedures involving contact with high-risk tissues (before guideline implementation).  

5.3. Dentistry

The risk of transmission of infection from dental instruments is thought to be very low provided optimal standards of infection prevention control and decontamination are maintained. However further guidance on the use of endodontic instruments is advised in the CMO and Chief Dental Officer’s letter of April 2008 (CMO (2007)5). Summary advice:

Endodontic reamers and files are treated as single-use. Single-use policies for these and other devices specified as single-use (e.g. matrix bands) are rigorously applied. Highest standards of decontamination are observed for all re-usable instruments. Manufacturers’ decontamination instructions are followed for all instruments and decontamination equipment.

For advice on infection control and CJD in dentistry contact Professor Andrew Smith by paging via Glasgow Royal Infirmary switchboard 0141 211 4000.

5.4. Intravascular injections

Plan care in advance where possible and as usual avoid sharps injuries and other parenteral exposures. Standard infection control precautions apply.

Always ensure the safe disposal of sharps and the safe disposal of clinical waste.
5.5. **Invasive procedures**

Ensure all procedures involving access to the subarachnoid space are undertaken with “single-use” instruments only, e.g. lumbar puncture including access to lumbar or extra ventricular drains and neuroradiological procedures. These should only be undertaken by trained staff who are fully aware of the risks.

5.5.1. **Procedure for Lumbar Puncture**

Plan the procedure in advance:
- single-use equipment only
- full PPE; gloves, apron and face protection
- place an incontinence pad underneath the patient to protect the sheet
- carefully decant the CSF into the container
- take care not to contaminate the outside of the container
- secure the specimen lid and place in a specimen bag
- used incontinence pad and PPE should be disposed of as clinical waste

The National CJD Unit and theatre staff must be informed in advance of any proposed biopsy request on high-risk tissue. These samples must be marked “DANGER of INFECTION”. Ensure theatre staff are aware of whether the specimen needs to be fixed or frozen for CJD.

Certain procedures involve contact with high-risk tissues and therefore additional precautions need to be taken to prevent cross-infection.

5.5.2. **Endoscopy**

**Full detailed Guidance on all endoscopic procedures can be found within Annex F:**


It is important that the full guidance is checked and adhered to. The following is a summary only.

The guidance given by the Medical Devices Agency (MDA) 2002(05) should be followed for all endoscopic procedures.
For all procedures:

- Channel cleaning brushes and, if biopsy forceps or other accessories have been passed, the rubber valve on the endoscope biopsy/ instrument channel port should be disposed of as clinical waste after each use.
- Single-use (i.e. disposable) biopsy forceps should be used routinely in all patients.
- Aldehyde disinfectants with fixative qualities tend to stabilise rather than inactivate prions and should not be used.

To decrease the risk of transmission of TSEs through endoscopic procedures, additional precautions for the decontamination of flexible endoscopes used in all patients with definite, probable or possible CJD/vCJD, and in those identified as “at increased risk” of developing CJD/vCJD. It is important that Annexe F of the CJD guidance is adhered to:


The main points include:

- Full traceability of the endoscope and any additional non-disposable devices used during the procedure.
- Where possible any cleaning tools and additional devices should be of single-use and discarded as clinical waste after the procedure.

For non neuro endoscopy on a person identified as at risk via the single question, for procedures involving contact with medium risk tissue, further information should be sought in the first instance. Infection Control should be contacted, if available and there is time, to conduct a full risk assessment. Generally the risk assessment takes account of the type of CJD/ vCJD of which the patient is at risk; the types of tissue involved and, in some cases, whether the procedure is invasive. The result of the risk assessment will determine whether special precautions should be used. If a risk is confirmed the instruments used should be either i) single use, ii) quarantined after use if re-usable (and destroyed if the risk is confirmed) or, rarely, iii) kept for exclusive use on the same patient. If the patient answers yes and Infection Control are not available, or time is of the essence, adopt a precautionary approach and quarantine all re-usable instruments until the full risk assessment can be undertaken. If the patient is unable to respond the procedure should go ahead using normal infection control precautions unless there is other information available on the patient’s risk status, e.g. GP letter, or if there is contact with high-risk tissues. In these situations re-usable instruments should be quarantined until the risk is confirmed.
Table 1: Endoscopy: CJD other than vCJD (adapted from Annexe F)

<table>
<thead>
<tr>
<th>Tissue Infectivity</th>
<th>Status of Patient</th>
<th>Symptomatic</th>
<th>Asymptomatic</th>
</tr>
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<tbody>
<tr>
<td></td>
<td></td>
<td>Definite/ Probable</td>
<td>Possible / Diagnosis Unclear¹</td>
</tr>
<tr>
<td>High</td>
<td></td>
<td>Single-use or destroy after use or quarantine³ for re-use exclusively on the same index patient</td>
<td>Single-use or quarantine pending diagnosis</td>
</tr>
<tr>
<td>Brain Spinal cord</td>
<td></td>
<td>Single-use or destroy after use or quarantine³ for re-use exclusively on the same index patient</td>
<td>Single-use or quarantine pending diagnosis</td>
</tr>
<tr>
<td>Medium</td>
<td>Olfactory Epithelium*</td>
<td>Single-use or destroy after use or quarantine³ for re-use exclusively on the same index patient</td>
<td>Single-use or quarantine pending diagnosis</td>
</tr>
<tr>
<td>Low/ none detectable</td>
<td>All other tissues</td>
<td>No special precautions</td>
<td>No special precautions</td>
</tr>
</tbody>
</table>

Notes

* The advice of the consultant carrying out the endoscopic procedure in the nasal cavity should be sought to determine whether a risk of contamination of the endoscope with olfactory epithelium can be excluded with confidence. If such contamination cannot be excluded, take precautions appropriate for medium infectivity tissues. See Annexe F for the full guidance for nasendoscopy.

1. This includes patients with neurological disease of unknown aetiology who do not fit the criteria for possible CJD but where a diagnosis of CJD is being actively considered (see also Annex B)

2. This advice refers to the use of flexible endoscopes in patients at risk of developing CJD. For guidance on the use of rigid endoscopes that can be autoclaved, refer to the guidance for the use of all surgical instruments in at risk patients in Part 4 of the CJD guidance.

3. Quarantined endoscopes may be re-used exclusively on the same individual patient if required. The principles behind the procedures recommended for quarantining of surgical instruments in Annex E of this Guidance should be followed except the endoscope should be fully cleaned and decontaminated immediately after use, before being quarantined. The endoscope should be decontaminated alone using an Automatic Endoscope Washer Disinfector (EWD). The EWD should be decontaminated as per paragraph F1 (e) of this guidance.

The most up-to-date version of this policy can be viewed at the following website: www.nhsggc.org.uk/infectionpreventionandcontrol
Table 2: Endoscopy: vCJD and CJD type uncertain (adapted from Annexe F)

<table>
<thead>
<tr>
<th>Tissue Infectivity</th>
<th>Status of Patient</th>
<th>Symptomatic</th>
<th>Asymptomatic</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Definite/ probable</td>
<td>Possible vCJD, possible sCJD or diagnosis Unclear(^1)</td>
<td>At risk (blood recipient from a donor who later developed vCJD)(^{***})</td>
</tr>
<tr>
<td>High Brain Spinal cord</td>
<td>Single-use or Destroy after use or quarantine(^3) for the re-use exclusively on the same patient.</td>
<td>Single-use Quarantine pending diagnosis</td>
<td>Single-use or Destroy after use or quarantine(^3) for the re-use exclusively on the same patient.</td>
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<td>Medium Olfactory Epithelium*</td>
<td>Single-use or Destroy after use or quarantine(^3) for the re-use exclusively on the same patient.</td>
<td>Single-use instruments or Quarantine pending diagnosis</td>
<td>Single-use or Destroy after use or quarantine(^3) for the re-use exclusively on the same patient.</td>
</tr>
<tr>
<td>Medium Lymphoid Tissue**</td>
<td>Single-use or Destroy after use or quarantine(^3) for the re-use exclusively on the same patient.</td>
<td>Single-use instruments or Quarantine pending diagnosis</td>
<td>No special precautions unless procedure is invasive(^1) If invasive use Single-use or Destroy after use or quarantine(^3) for the re-use exclusively on the same patient.</td>
</tr>
<tr>
<td>Low All other tissues</td>
<td>No special precautions</td>
<td>No special precautions</td>
<td>No special precautions</td>
</tr>
</tbody>
</table>

Notes

* The advice of the consultant carrying out the endoscopic procedure in the nasal cavity should be sought to determine whether a risk of contamination of the endoscope with olfactory epithelium can be excluded with confidence. If such contamination cannot be excluded, take precautions appropriate for medium infectivity tissues. See Annexe F for the full nasendoscopy guidance.

** For the purposes of this Annex (F), lymphoid tissue refers to the spleen, thymus, tonsils and adenoids, lymph nodes, the appendix and the gastro-intestinal tract sub-mucosa.

*** A small number of individuals are known to have received labile blood components (whole blood, red cells, white cells or platelets) from a donor who later developed vCJD.

The most up-to-date version of this policy can be viewed at the following website: [www.nhsggc.org.uk/infectionpreventionandcontrol](http://www.nhsggc.org.uk/infectionpreventionandcontrol)
1 This includes patients with neurological disease of unknown aetiology who do not fit the criteria for possible vCJD but where a diagnosis of vCJD is being actively considered (see also Annex B of the guidance).

2 This advice refers to the use of flexible endoscopes in patients at risk of developing vCJD. For guidance on the use of rigid endoscopes that can be autoclaved, refer to the guidance for the use of all surgical instruments in at risk patients in Part 4 of the guidance.

3 Quarantined endoscopes may be re-used exclusively on the same individual patient if required. The principles behind the procedures recommended for quarantining of surgical instruments in Annex E of this Guidance should be followed except the endoscope should be fully cleaned and decontaminated immediately after use, before being quarantined. The endoscope should be decontaminated alone using an Automatic Endoscope Washer Disinfector (EWD). The EWD should be decontaminated as per paragraph F1 (e) of this guidance.

4. For Asymptomatic patients “at increased risk” through receipt of labile blood components (whole blood, red cells, white cells or platelets) from a donor who later developed vCJD. For non neuro and non nasal endoscopies providing decontamination of the endoscope is to approved standards, the use of the instrument for inspection in the absence of an invasive procedure, see Table F2b of Annexe F, is deemed to be a low risk procedure. If an invasive procedure is carried out, the possibility of contamination of the instrument channel with lymphoid tissue means the endoscope should be decontaminated singly as at F1(c-f), then quarantined pending assessment of likely contact with potentially infected tissue. If this is considered possible the endoscope should be removed from use.

5.6. Surgical Instruments

- Tonsillectomies must be undertaken using SINGLE-USE EQUIPMENT ONLY regardless of CJD status.
- Any re-usable instrument in contact with adeno-tonsillar tissue from a suspected vCJD case or a patient at risk of vCJD should be quarantined until a definitive diagnosis is made or the risk status established and then either permanently removed (if positive vCJD diagnosis, or risk status) or reprocessed according to “best practice” (if definite alternative diagnosis is made or risk status is not established). Instruments used on known cases must be destroyed.
- Use of diathermy in tonsillectomy is restricted to situations where it is clinically necessary.
- Use single-use biopsy instruments wherever possible.
- Wherever possible use single-use equipment, disposable tips and protective instrument sheaths.
- All other instruments used in adeno-tonsillar surgery should be decontaminated according to current best practice for ‘medium risk’ tissues.

Ensure equipment with a lumen is irrigated as soon as possible after surgery to facilitate adequate decontamination. Also see full endoscope guidance as given above.

The most up-to-date version of this policy can be viewed at the following website: www.nhsggc.org.uk/infectionpreventionandcontrol
The most up-to-date version of this policy can be viewed at the following website:

www.nhsggc.org.uk/infectionpreventionandcontrol

For general surgery Annexe M, **Managing v CJD risk in General Surgery and liver transplantation**

[https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/209780/Annex_M_-_Mananging_vCJD_risk.pdf](https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/209780/Annex_M_-_Mananging_vCJD_risk.pdf) gives advice on handling instruments that come into contact with medium risk tissues involved in liver transplants and general surgical procedures. It applies to all patients with or at risk of variant CJD undergoing these procedures. For general surgery medium risk tissues include: gut-associated lymphoid tissue, lymph nodes and other organised lymphoid tissues containing follicular structures, appendix, spleen, thymus and tonsil. The guidance applies to surgical procedures where instruments come into contact with the cut surface of such tissues, eg operations on the bowel, porta hepatis or cervical lymph nodes. It does not apply to procedures where only lymphatic channels are cut.

Further, it applies to patients who have been identified as at risk of variant CJD or those who are symptomatic with probable or definite variant CJD. It does not apply to patients with or at risk of other prion diseases such as familial CJD and sporadic or iatrogenic CJD that is not variant CJD.

5.7. **Quarantining Instruments**

A secure dedicated storage box must be used if instruments are to be quarantined. Clear indication as the identity of the patient must remain with the instruments. Quarantined instruments are normally kept until a definite diagnosis is made or risk status confirmed or refuted, or in some situations, if the instruments are to be used on the same patient at a near future date. If a definite alternative diagnosis is made or the risk status is refuted then the instruments may be put back into circulation after “best practice” decontamination. Full explanation and detailed instructions to be found within Annex E **HOWEVER PLEASE NOTE** in NHS Scotland due to the Glennie Technical Requirements instruments should NOT be cleaned prior to quarantining via CDU but should be wiped down with clean water to remove any blood or tissue and dried prior to quarantining.


The decision to destroy surgical instruments will be taken by the local IPCT and the Manager of the Central Decontamination Unit.
6. **Occupational Exposure incidents from patients thought to have definite, probable or possible CJD.**

Avoid sharp injuries and other forms of parenteral exposure, and ensure the safe disposal of sharps and contaminated waste. If incidents occur:

- Gently encourage wounds to bleed.
- Gently wash with warm soapy water – avoid scrubbing.
- Rinse, dry and cover with a waterproof dressing.
- Splashes into the eyes or mouth should be dealt with by thorough irrigation.

All exposures must be reported using incident reporting systems (Datix) and subsequently under RIDDOR if medically diagnosed.

7. **General Healthcare**

7.1. **Nursing procedures**

Normal nursing care should be given to all patients with CJD/ TSE. There is no requirement to isolate these patients. Epidemiological evidence supports general standard infection control precautions to be satisfactory and posing no risk to staff or relatives.

7.2. **Bed linen and patient’s clothing**

No special precautions are needed local policies apply.

7.3. **Visitors**

No restrictions.

7.4. **Childbirth**

Standard infection control precautions apply.

7.5. **Transfer of patients to other wards / healthcare settings**

No restrictions but ensure those receiving the patient are aware of any established risk.

7.6. **Terminal clean**

Follow NHSGGC SOP for Terminal Clean of an Isolation Room. For further information see Part 4:

7.7. **Procedures after Death**

Follow normal procedures but ensure that the mortuary card states ‘Infection Risk’ and is adhered to the body bag. The mortuary and funeral directors must also be informed in line with NHSGGC Last Offices SOP.

**Relatives**
- Relatives may view and have superficial contact with the body.
- Viewing and kissing the body is not restricted.
- No extra precautions need be taken for burial or cremation, either of which is acceptable.

**Undertakers and embalmers**
- Handling of intact bodies poses no additional risk to the undertaker.
- Contact should be minimised to prevent penetrating injuries.
- Cosmetic work may be performed as normal.
- Embalming procedures should be avoided.

7.8. **Post mortems**

For further details please see Annex H:

7.9. **Donation of Organs**

Not recommended from any patient with **definite, probable or possible** CJD.

7.10. **Public health advice on how to stop CJD spreading to other people**

This advice is for people who have been identified as being at increased risk of CJD. To reduce the risk of spreading CJD to other people please follow this advice:
- Do no 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.
- Do not 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.
- You are advised to 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 most up-to-date version of this policy can be viewed at the following website:
[www.nhsggc.org.uk/infectionpreventionandcontrol](http://www.nhsggc.org.uk/infectionpreventionandcontrol)
8. **Laboratory Investigations**

All laboratories in the UK have procedures in place to ensure cross-contamination of samples is prevented. It is essential that "high-risk" stickers are placed on samples from patients with known or suspected CJD and that the laboratory is telephoned in advance of sending the sample.

Specimens from patients with **definite, probable** or **possible** CJD or asymptomatic patients at risk from CJD should be treated as follows:

- Non-lymphoid and non-neurological specimens, e.g. **blood**, **urine**, **faeces**, **swabs**, etc., can be collected, processed and handled as for any other patient.
- CSF samples are designated low risk. Normal standard infection control precautions should be taken for biochemical analysis.
- Use disposable equipment where practicable.
- Items contaminated by the specimens should be destroyed by incineration, or, decontaminated as in the spillages section.
- If automated equipment is used, ensure decontamination takes place prior to servicing.
- Microscopes must be cleaned and regularly maintained to prevent the accumulation of potentially contaminated debris.


9. **Training and Education for Healthcare Workers (HCW)**

All HCWs caring for patients with **definite, probable** or **possible** CJD must have received education on Standard Precautions and be aware of this policy. All HCWs undertaking biopsy, blood and lumbar puncture samples from patients with **definite, probable** or **possible** CJD, or from those at risk of CJD, must be trained to do so and be aware of the CJD Policy and also be aware of the need to use single-use equipment.

The most up-to-date version of this policy can be viewed at the following website: [www.nhsggc.org.uk/infectionpreventionandcontrol](http://www.nhsggc.org.uk/infectionpreventionandcontrol)
10. References


The National Creutzfeldt-Jakob Disease Surveillance Unit (personal communication)

Medical Devices Agency Decontamination of Endoscopes July 2002 MDA DB (2002)05


National Institute for Health and Clinical Excellence: patient safety and reduction of risk of transmission of Creutzfeldt-Jakob disease (CJD via interventional procedures. November 2006 (Guidance endorsed by NHS QIS for implementation by NHS Scotland)

“Endoscopy and individuals at risk of vCJD for public health purposes” A consensus statement from the British Society of Gastroenterology Decontamination Working Group and the ACDP TSE Working Group Endoscopy and vCJD Sub-Group (November 2005)

Bibliography


11. Websites

https://www.gov.uk/search?q=tse&tab=government-results
http://www.cjd.ed.ac.uk/

The most up-to-date version of this policy can be viewed at the following website: www.nhsggc.org.uk/infectionpreventionandcontrol
Appendix 1 – CJD Risk Assessment

a) Introduction

The following risk assessment includes a generic form and associated algorithm for administering the single question and/or the single question and at risk question and, as appropriate, questions relevant for implementation of the NICE guidance.

The clinician undertaking the pre-surgery assessment should also:

- Check the patient’s medical notes and/or referral letter for any mention of CJD or vCJD status.
- Consider whether there is a risk that the patient may be showing the early signs of CJD or vCJD.

b) CJD Risk Assessment

<table>
<thead>
<tr>
<th>Name</th>
<th>Address Use Label</th>
<th>CHI number</th>
<th>Date</th>
</tr>
</thead>
<tbody>
<tr>
<td>CJD is a very rare, rapidly progressing disease of the nervous system that is caused by an abnormal protein (a prion). It is only transmissible if tissue (mainly the brain) from an infected person or animal enters the body of another. CJD is not spread by social contact, coughing, sneezing, kissing, sexual intercourse, childbirth or breast-feeding. Doctors, nurses, relatives and others caring for patients with CJD are not at risk of contracting CJD.</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

To reduce the chance of spreading CJD we need to identify patients whose risk of developing CJD may be increased. The treatment of such patients will not be affected in any way, but appropriate measures to prevent transmission to other patients via surgical instruments will be taken.

How to use this flow sheet:

CJD risk assessment should be performed for all patients undergoing endoscopy or surgery. This applies to both elective and emergency procedures. When it is not possible to assess the risk in advance, (e.g. before emergency procedures on very ill or unconscious patients), for procedures involving contact with high-risk tissues, re-usable instruments should be quarantined pending obtaining the relevant information from the patient’s relatives or General Practitioner. For invasive procedures which do not involve contact with high-risk tissues only the single question is asked. In this situation when the patient is unable to respond to the single question and no other information is available the procedure should go ahead using standard infection control precautions. If the patient says yes to the single question but infection control are not available to conduct the full risk assessment the procedure should go ahead and re-usable instruments should be quarantined pending full risk assessment as soon as possible.
c) Invasive procedure involving contact with low or medium risk tissue only:

**Ask the Single question: Responses and associated actions**

<table>
<thead>
<tr>
<th>Question: Have you been notified that you are at risk of CJD for public health purposes?</th>
<th>Patient’s Response</th>
<th>Action</th>
</tr>
</thead>
<tbody>
<tr>
<td>No</td>
<td>Surgery or endoscopy should proceed using standard infection control procedures unless the procedure is likely to lead to contact with high-risk tissue.</td>
<td></td>
</tr>
</tbody>
</table>
| Yes | Ask the patient to explain further. Contact infection control, if available and there is time, to conduct a full risk assessment.  
For an invasive procedure on a person identified as at risk via the single question, for procedures involving contact with medium risk tissue, further information should be sought in the first instance.  
Generally the risk assessment takes account of the type of CJD/ vCJD of which the patient is at risk; the types of tissue involved and, in some cases, whether the procedure is invasive. The result of the risk assessment will determine whether special precautions should be used.  
If a risk is confirmed the instruments used should be either i) single use, ii) quarantined after use if re-usable (and destroyed if the risk is confirmed) or, rarely, iii) kept for exclusive use on the same patient (most commonly endoscopes) in line with the national CJD guidance (generally Annexe F and Annexe M).  
If the risk is not confirmed they should return to circulation after normal decontamination procedures and a check of the integrity of the instrument.  
If the patient answers yes to the single question and infection control is not available, or time is of the essence, the procedure should go ahead and adopt a precautionary approach and quarantine all re-usable instruments until the full risk assessment can be undertaken.  
Other actions and public health advice for those patients where the risk is confirmed:  
• Contact infection control (who will contact PHPU) (PHPU will inform HPS for surveillance purposes).  
• Infection Control or Public Health will discuss the risk with the patient (using patient information leaflet) and give the patient the usual infection control and public health advice. The patient should be advised:  
  • To inform healthcare staff if they need to undergo an invasive surgical, medical or dental procedure;  
  • To inform a family member or someone close to them, in case they need emergency surgery or endoscopy in the future.  
  • Not to donate blood, organs or tissues, including bone marrow, sperm, eggs or breast milk.  
  • The patient’s GP should be informed and advised, record the patient’s CJD risk status in their primary care records. The GP should also include this information in any referral letter should the patient require invasive surgical, medical or dental procedures.  
• Infection Control will inform medical records so that the electronic records can be flagged. Infection Control and Public Health will work together to identify any previous procedures which require to be reported to HPS. |
| Unable to respond | Surgery or endoscopy should proceed using standard infection control procedures unless the procedure is likely to lead to contact with high-risk tissue. |
d) Invasive procedure involving contact with high-risk tissues:

The single question and at risk questions:

<table>
<thead>
<tr>
<th>Question</th>
<th>Notes</th>
<th>Response</th>
<th>Actions, if yes or unable to respond</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Single question Have you been notified that you are at risk of CJD for public health purposes?</td>
<td>Patients should be considered to be at risk from genetic forms of CJD if they have or have had: i) Genetic testing, which has indicated that they are at significant risk of developing CJD or other prion disease. ii) A blood relative known to have a genetic mutation indicative of genetic CJD or other prion disease. iii) Two or more blood relatives affected by CJD or other prion disease.</td>
<td>If yes to any of the questions or unable to respond (and insufficient time to obtain the information from family or GP).</td>
<td>The instruments used should be either i) single use, or ii) quarantined after use if re-usable or, rarely, iii) kept for exclusive use on the same patient. For further details See Annex J at: <a href="https://www.gov.uk/government/publications/guidance-from-the-acdp-tse-risk-management-subgroup-formerly-tse-working-group">https://www.gov.uk/government/publications/guidance-from-the-acdp-tse-risk-management-subgroup-formerly-tse-working-group</a></td>
</tr>
<tr>
<td>2. Have you a history of CJD or other prion disease in your family? If yes, please specify.</td>
<td>Recipients of hormone derived from human pituitary glands, e.g. growth hormone or gonadotrophin, have been identified as at increased risk of sporadic CJD. In the UK, the use of human-derived growth hormone was discontinued in 1985 but human-derived products may have continued to be used in other countries. In the UK, the use of human-derived gonadotrophin was discontinued in 1973 but may have continued in other countries after this time.</td>
<td>Follow actions as stated.</td>
<td>If this risk is confirmed quarantined re-usable instruments require to be destroyed. If it is not confirmed they should return to circulation after normal decontamination procedures and a check of the integrity of the instrument.</td>
</tr>
<tr>
<td>3. Have you ever received growth hormone or gonadotrophin treatment? If yes, please specify: i) whether the hormone was derived from human pituitary glands ii) the year of treatment , and iii)whether the treatment was received in the UK or in another country</td>
<td></td>
<td></td>
<td>For those patients where the risk is confirmed: ▪ Contact infection control (who will contact PHPU) (PHPU will inform HPS for surveillance purposes). ▪ Infection Control or Public Health will discuss the risk with the patient (using patient information leaflet) and give the patient the usual infection control and public health advice. The patient should be advised: o To inform healthcare staff if they need to undergo an invasive surgical, medical or dental procedure; o To inform a family member or someone</td>
</tr>
</tbody>
</table>
### 4. Have you ever had surgery on your brain or spinal cord?

(a) Individuals who underwent intradural brain or intradural spinal surgery before August 1992 who received (or might have received) a graft of human-derived dura mater are “at increased risk” of transmission of sporadic CJD (unless evidence can be provided that human-derived dura mater was not used).

(b) **NICE guidance** emphasises the need for a separate pool of new neuroendoscopes and reusable surgical instruments for high-risk procedures on children born since 1st January 1997 and who have not previously undergone high-risk procedures. These instruments and neuroendoscopes should not be used for patients born before 1st January 1997 or those who underwent high-risk procedures using reusable instruments before the implementation of this guidance.

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The patient’s GP should be informed and advised to record the patient’s CJD risk status in their primary care records. The GP should also include this information in any referral letter should the patient require invasive surgical, medical or dental procedures.

- **Infection Control** will inform medical records so that the electronic records can be flagged.
- Infection control and Public Health will work together to identify any previous procedures which require to be reported to HPS.

Patients who are at increased risk of genetic forms of CJD should be offered the opportunity of referral to the National Prion Clinic, based at the National Hospital for Neurology and Neurosurgery, Queen Square, London: [http://www.nationalprionclinic.org/](http://www.nationalprionclinic.org/) Patients who are at increased risk of sporadic CJD due to receipt of human-derived growth hormone or gonadotrophin should be offered the opportunity of referral to the UCL Institute of Child Health, London. Contact: L.Davidson@ich.ucl.ac.uk, 020 7404 0536

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The most up-to-date version of this policy can be viewed at the following website: [www.nhsggc.org.uk/infectionpreventionandcontrol](http://www.nhsggc.org.uk/infectionpreventionandcontrol)
e) CJD Algorithm

CJD Algorithm 1:

Invasive procedure involving contact with medium risk tissue (no contact with high risk tissue) for endoscopy see Tables 1 and 2 of this policy and annex F.

Ask Single Question: Have you been notified that you are at increased risk of CJD for public health purposes?
Also check if mentioned in referral letter or has electronic notes tagged.

No

Unable to respond

Proceed using normal infection control precautions

Yes

Ask the patient to explain further. Contact infection control, if available and there is time, to conduct a full risk assessment. Generally, the risk assessment takes account of the type of CJD/CJD of which the patient is at risk, the types of tissue involved and, in some cases, whether the procedure is invasive.

If infection control is not available, or time is of the essence, adopt a precautionary approach and quarantine all re-usable instruments until the full risk assessment can be undertaken.

Result of risk assessment

Procedures with potential contact with olfactory epithelium: for all at risk categories of CJD/CJD: ask the surgeon if there is to be contact with the olfactory epithelium. If yes, or there is any doubt, additional infection control measures are required as described below.

For general surgery follow Annexe M: generally, where a risk of variant CJD (vCJD) is identified and there is contact with medium risk tissue (including gut associated lymphoid tissue, lymph nodes and other organised lymphoid tissues containing follicular structures, appendix, spleen, thymus and tonsil) and surgical instruments will possibly come into contact with the cut surface of these tissues additional infection control measures are required as described below.

If risk is identified following the risk assessment: Use disposable instruments and/or quarantine reusable instruments.

In exceptional circumstances keep instruments for re-use exclusively on the same patient.

Confirm the risk status with the GP

If the risk is confirmed infection control will:
- Explain the risk to the patient
- Give public health advice
- Inform public health and the GP
- Write in the notes and ensure electronic file tagged.

If the risk is confirmed destroy any quarantined re-usable instruments or in exceptional circumstances keep for re-use exclusively on the same patient.

If the risk is not confirmed the quarantined instruments can be re-processed.
THE MANAGEMENT OF PATIENTS WITH TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHY (TSE) INCLUDING ALL FORMS OF Creutzfeldt-Jakob Disease (CJD)

The most up-to-date version of this policy can be viewed at the following website:

www.nhsggc.org.uk/infectionpreventionandcontrol
f) Sign Off page

Summary

(Please circle)

Patient may be at risk (see flow sheet for details)  Yes  No

Operation is on high-risk (CJD)/ high or medium risk (vCJD) tissues  Yes  No

Signed

Designation

Date
**Appendix 2 – Tissue Risk Categories**

**Key:**  
Positive = tested positive  
Negative = tested negative  
NT = not tested  
P = infectivity proven in experimental transmission studies

<table>
<thead>
<tr>
<th>Tissue</th>
<th>Presence of abnormal prion protein and level of infectivity</th>
<th>CJD other than vCJD</th>
<th>vCJD</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>PrP&lt;sup&gt;TSE&lt;/sup&gt; detected</td>
<td>Assumed level of infectivity</td>
<td>PrP&lt;sup&gt;TSE&lt;/sup&gt; detected</td>
</tr>
<tr>
<td>Brain</td>
<td>Positive</td>
<td>High P</td>
<td>Positive</td>
</tr>
<tr>
<td>Spinal cord</td>
<td>Positive</td>
<td>High P</td>
<td>Positive</td>
</tr>
<tr>
<td>Cranial nerves, specifically the entire optic nerve and only the intracranial components of the other cranial nerves</td>
<td>Positive</td>
<td>High</td>
<td>Positive</td>
</tr>
<tr>
<td>Cranial ganglia</td>
<td>Positive</td>
<td>High</td>
<td>Positive</td>
</tr>
<tr>
<td>Posterior eye, specifically the posterior hyaloid face, retina, retinal pigment epithelium, choroid, subretinal fluid, optic nerve</td>
<td>Positive</td>
<td>High P</td>
<td>Positive</td>
</tr>
<tr>
<td>Pituitary gland</td>
<td>Positive</td>
<td>High (?)</td>
<td>Positive</td>
</tr>
<tr>
<td>Spinal ganglia&lt;sup&gt;1&lt;/sup&gt;</td>
<td>Positive</td>
<td>Medium</td>
<td>Positive</td>
</tr>
<tr>
<td>Olfactory epithelium</td>
<td>Positive</td>
<td>Medium</td>
<td>NT</td>
</tr>
<tr>
<td>Dura mater&lt;sup&gt;2&lt;/sup&gt;</td>
<td>Negative</td>
<td>Low</td>
<td>Positive</td>
</tr>
<tr>
<td>Tonsil</td>
<td>Negative</td>
<td>Low</td>
<td>Positive</td>
</tr>
<tr>
<td>Lymph nodes and other organised lymphoid tissues containing follicular structures</td>
<td>Negative</td>
<td>Low P</td>
<td>Positive</td>
</tr>
<tr>
<td>Gut-associated lymphoid tissue</td>
<td>Negative</td>
<td>Low</td>
<td>Positive</td>
</tr>
<tr>
<td>Appendix</td>
<td>Negative</td>
<td>Low</td>
<td>Positive</td>
</tr>
<tr>
<td>Spleen</td>
<td>Positive</td>
<td>Low P</td>
<td>Positive</td>
</tr>
<tr>
<td>Thymus</td>
<td>Negative</td>
<td>Low</td>
<td>Positive</td>
</tr>
<tr>
<td>Anterior eye and cornea</td>
<td>Negative</td>
<td>Low</td>
<td>Negative</td>
</tr>
<tr>
<td>Peripheral nerve</td>
<td>Positive</td>
<td>Low</td>
<td>Positive</td>
</tr>
<tr>
<td>Skeletal muscle</td>
<td>Positive</td>
<td>Low</td>
<td>Positive</td>
</tr>
<tr>
<td>Dental Pulp</td>
<td>Negative</td>
<td>Low</td>
<td>Negative</td>
</tr>
<tr>
<td>Gingival Tissue</td>
<td>NT</td>
<td>Low</td>
<td>Negative</td>
</tr>
<tr>
<td>Blood and bone marrow</td>
<td>NT</td>
<td>Low</td>
<td>Negative</td>
</tr>
<tr>
<td>CSF&lt;sup&gt;3&lt;/sup&gt;</td>
<td>Negative</td>
<td>Low P</td>
<td>Negative</td>
</tr>
<tr>
<td>Placenta</td>
<td>Negative</td>
<td>Low</td>
<td>Negative</td>
</tr>
<tr>
<td>Urine</td>
<td>Negative</td>
<td>Low</td>
<td>Negative</td>
</tr>
<tr>
<td>Other tissues</td>
<td>Negative</td>
<td>Low P</td>
<td>Positive</td>
</tr>
</tbody>
</table>

---

1. Spinal ganglia have a high assumed level of infectivity in the WHO Guidelines. However, unpublished results on the infectivity of spinal ganglia indicate that this tissue is of medium infectivity.

2. Dura mater is designated low infectivity as virtually no detectable abnormal prion protein has been found in cases of CJD; however, as grafts of these tissues are associated with CJD transmission, probably as a result of contamination by brain and because of the lengthy period of implantation in the CNS, procedures conducted on intradural tissues (i.e. brain, spinal cord and intracranial sections of cranial nerves) or procedures in which human dura mater was implanted in a patient prior to 1992, remain high-risk.

3. Although PrP<sup>TSE</sup> has not been detected in the CSF in either sporadic or variant CJD (15), experimental transmission of infectivity has been achieved from CSF in sporadic CJD in 4 of 27 primates by intracerebral inoculation (9) indicating that levels of infectivity are likely to be much lower than in the central nervous system.

4. PrP<sup>TSE</sup> has been detected in dura mater, skin, kidney, liver, pancreas, ovary and uterus in a case of vCJD in USA with a lengthy duration of illness (16). Earlier studies of these tissues in UK vCJD cases gave negative results (2, 8, 17).
Appendix 3 – Diagnostic Criteria

SPORADIC CJD

A) DEFINITE: Confirmed by neuropathological or immunocytochemical confirmation only.

B) PROBABLE: Rapidly progressive dementia and at least two of the following four symptoms:
   (a) Myoclonus
   (b) Visual or cerebellar problems
   (c) Pyramidal or extra pyramidal features
   (d) Akinetic mutism

Plus typical EEG with generalised triphasic periodic complexes at approximately 1 second,

or clinical criteria for possible vCJD and a positive assay for 14-3-3 protein in CSF.

C) POSSIBLE Rapidly progressive dementia, with two of the symptoms listed in B) a-d above and duration of less than 2 years.

IATROGENIC CJD
Progressive Cerebellar Syndrome in a pituitary hormone recipient or sporadic CJD with a recognised exposure risk (e.g. dura mater transplant). A definite diagnosis for iatrogenic requires neuropathological confirmation.

FAMILIAL CJD
Patients will have Definite or Probable CJD plus a first degree relative will also have had Definite or Probable CJD or a neuropsychiatric disorder plus a disease-specific mutation in the prion protein gene.

VARIANT CJD
DEFINITE: Progressive neuropsychiatry disorder and neuropathological confirmation of the disease, showing spongiform change and extensive PrP-c deposition with florid plaques throughout the cerebrum and cerebellum.
PROBABLE: Progressive neuropsychiatric disorder of duration greater than 6 months where routine investigations do not suggest an alternative diagnosis with at least 4 of the following symptoms:

(a) early psychiatric symptoms (depression, anxiety, apathy, withdrawal, delusions)
(b) persistent painful sensory symptoms (including both frank pain and/or unpleasant dysaesthesia)
(c) ataxia
(d) myclonus or chorea or dystonia
(e) dementia

EEG does not show changes as in sCJD but there is a symmetrical high signal in the posterior thalamus on a MRI brain scan.

There will be no history of potential iatrogenic exposure.

1. Progressive neuropsychiatric disorder for a period of longer than 6 months, where routine investigations do not support an alternative diagnosis and where there is no history of potential iatrogenic exposure plus a positive tonsil biopsy for PrP – res.

POSSIBLE vCJD: Progressive neuropsychiatric disorder for a period of longer than 6 months, where routine investigations do not support an alternative diagnosis and where there is no history of potential iatrogenic exposure. They will have at least 4 of the 5 symptoms listed above and an EEG does not show the typical appearance of sCJD or no EEG has been performed.
Appendix 4: Categorisation of patients at risk

<table>
<thead>
<tr>
<th>Patient Groups</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptomatic patients</td>
<td>Patients who fulfil the diagnostic criteria for definite, probable or possible CJD or variant CJD. Patients with neurological disease of unknown aetiology who do not fit the criteria for possible CJD or vCJD, but where the diagnosis of CJD is being actively considered.</td>
</tr>
<tr>
<td>Patients at increased risk from genetic forms of CJD</td>
<td>Individuals who have been shown by specific genetic testing to be at significant risk of developing CJD. Individuals who have a blood relative known to have a genetic mutation indicative of genetic CJD. Individuals who have or have had two or more blood relatives affected by CJD or other prion disease.</td>
</tr>
<tr>
<td>Patients identified as at increased risk of variant CJD through receipt of blood from a donor who later developed variant CJD</td>
<td>Individuals who have received labile blood components (whole blood, red cells, white cells or platelets) from a donor who later went on to develop variant CJD.</td>
</tr>
<tr>
<td>Patients at increased risk of CJD/variant CJD through iatrogenic exposures</td>
<td>Recipients of hormone derived from human pituitary glands eg growth hormone, gonadotrophin are at increased risk of sporadic CJD. In the UK the use of human derived gonadotrophin was discontinued in 1973, and use of cadaver-derived human growth hormone was banned in 1985. However the use of human derived products may have continued in other countries after these dates. Individuals who underwent intradural brain or intraspinal surgery before August 1992 who received (or might have received) a graft of human derived dura mater are at increased risk of transmission of sporadic CJD (unless evidence can be provided that human-derived dura mater was not used). Individuals who have had surgery using instruments that had been used on someone who went on to develop CJD/vCJD or was at increased risk of CJD/vCJD.</td>
</tr>
<tr>
<td></td>
<td>Individuals who have been identified as having received blood or blood components from 300 or more donors since January 1990. Individuals who have given blood to someone who went on to develop variant CJD. Individuals who have received blood from someone who has also given blood to a patient who went on to develop variant CJD. Individuals who have been treated with certain implicated UK sourced plasma products between 1990 and 2001 (inclusive).</td>
</tr>
</tbody>
</table>

Recipients of ocular transplants, including corneal transplants, are not considered to be “at increased risk” of CJD/vCJD.