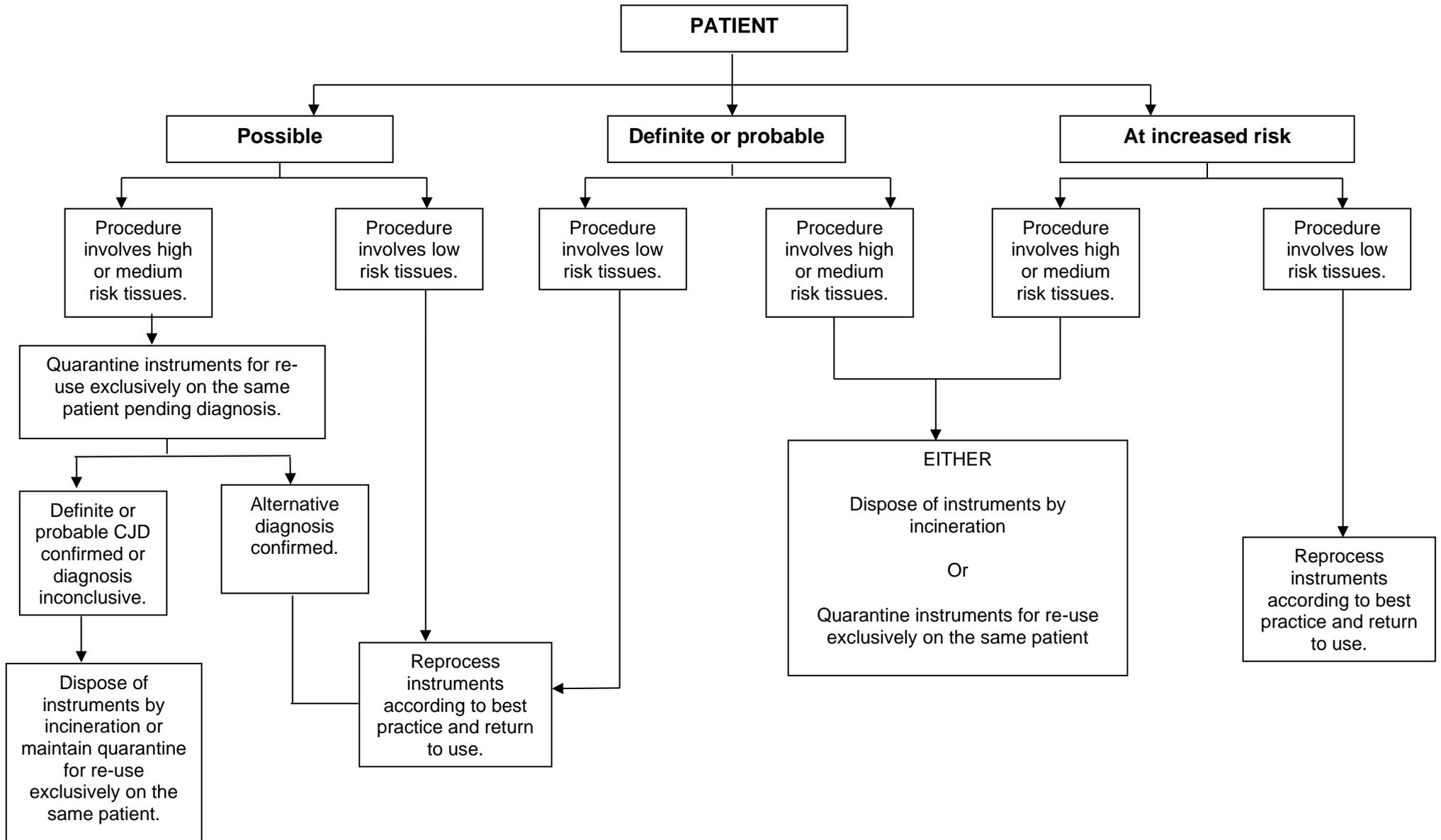


# **Patients who are Symptomatic or at increased risk of transmissible Creutzfeldt–Jakob disease (CJD) Policy**

**V2.0**

**June 2023**

# Summary



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### **Data Protection Act 2018 (General Data Protection Regulation – GDPR) Legislation**

The Trust has a duty under the Data Protection Act 2018 and General Data Protection Regulations 2016/679 to ensure that there is a valid legal basis to process personal and sensitive data. The legal basis for processing must be identified and documented before the processing begins. In many cases we may need consent; this must be explicit, informed, and documented. We cannot rely on opt out, it must be opt in.

Data Protection Act 2018 and General Data Protection Regulations 2016/679 is applicable to all staff; this includes those working as contractors and providers of services.

For more information about your obligations under the Data Protection Act 2018 and General Data Protection Regulations 2016/679 please see the Information Use Framework Policy or contact the Information Governance Team

Royal Cornwall Hospital Trust      [rch-tr.infogov@nhs.net](mailto:rch-tr.infogov@nhs.net)

## **1. Introduction**

- 1.1. The prevention of transmission of infection and the provision of safe instruments are fundamental for patient care. Incorrect procedures for the handling or processing of instruments used on patients infected with any form of Transmissible Spongiform Encephalopathy (TSE), namely Creutzfeldt - Jakob disease (CJD) or Variant Creutzfeldt-Jakob Disease (vCJD) can present an infection risk to patients on whom instruments are subsequently used.
- 1.2. This policy describes the steps, which must be taken to manage patient treatment in order to minimise risk and is based on national guidelines for preventing the transmission of TSEs.
- 1.3. This version supersedes any previous versions of this document.

## **2. Purpose of this Policy/Procedure**

The purpose of this protocol is to provide guidance to staff on the precautions necessary to minimise the risk of occupational exposure to CJD and to prevent transmission of CJD between patients.

## **3. Scope**

This policy applies to all staff working for or on behalf of the Royal Cornwall Hospitals NHS Trust including volunteer, temporary, locum, bank, agency and contracted staff.

## **4. Definitions / Glossary**

Definitions are contained within the text.

## **5. Ownership and Responsibilities**

### **5.1. Role of the Chief Executive**

The Chief Executive Officer (CEO) is responsible for ensuring that there are effective arrangements for infection prevention and control within the Trust. This includes determining the mechanisms by which the Trust Board ensures that there are adequate resources available to secure effective prevention and control of healthcare associated infections.

### **5.2. Role of the Director of Infection Prevention and Control (DIPC)**

The DIPC is responsible for overseeing the implementation of the policy, for reporting any concerns and performance in infection prevention and control to the Trust Board and CEO.

### **5.3. Role of Associate Directors**

Associate Directors:

- Will ensure all staff responsible for the management of patients with CJD comply with the policy and procedures laid down in this document and have the necessary resources.
- Will ensure their staff receive appropriate support and training in the management of CJD and vCJD.

### **5.4. Role of the Consultant responsible for the Patient**

The Consultant responsible for the patient will:

- Ensure that the Infection Prevention and Control Team are informed of any patient in any of the above categories of CJD & vCJD.
- Ensure appropriate departments are informed before any invasive procedures are carried out.
- Ensure that any case where TSE/CJD & vCJD of any type is a possible diagnosis is reported to the National CJD Research and Surveillance Unit (CJDSU) (Tel: 0131 537 2128) the National Prion Clinic London (Tel: 0203 448 4037) and the local UKHSA health protection team (0300 303 8162) so that necessary action can be taken particularly with regards to making or excluding the diagnosis.

### **5.5. Role of the Infection Prevention and Control Team**

The Infection Prevention and Control Team will:

- Provide guidance and support on the practical application of this protocol.
- Monitor, evaluate and review the policy in the light of new evidence.

### **5.6. Role of the Occupational Health Department**

The Occupational Health Department Record any significant exposure to Health Care Workers from blood or CSF & other source.

### **5.7. Role of the Infection Prevention and Control Steering Group**

The IPAC Steering Group is responsible for the implementation and monitoring of this policy.

### **5.8. Role of the Hospital Infection Prevention and Control Committee**

The Hospital Infection Prevention and Control Committee are responsible for approving this document.

## 5.9. Role of All Staff Members

All staff members working on Trust premises, including Trust employed staff, contractor staff, agency and locum staff are responsible for:

- Adhering to this policy.
- Reporting breaches of this policy to the person in charge and to their line manager.

## 5.10. Role of Consultant Medical Staff

Consultant Medical staff are responsible for ensuring their junior staff read and understand this policy and adhere to the principles contained in it at all times.

## 5.11. Role of Managers

Line Managers are responsible for monitoring implementation of this policy, and understanding this policy, and adhering to the principles contained in it at all times.

# 6. Standards and Practice

## 6.1. Background

TSE's, otherwise known as prion diseases, are rare, fatal, degenerative diseases affecting the central nervous system, that occur in humans and other mammals.

The human TSE's occur in three groups:

- Idiopathic diseases: sporadic CJD and sporadic fatal insomnia.
- Familial diseases: familial CJD, Gerstmann-Straussler-Scheinker Syndrome (GSS) Fatal Familial Insomnia (FFI).
- Acquired diseases:
  - Human agents: Kuru and iatrogenic CJD.
  - Bovine agent: variant CJD

All human TSE's are very rare; the worldwide incidence of CJD is about 1 per million people each year. Sporadic CJD accounts for around 85% of all human TSE's: familial TSE's account for around 10-15% of cases and the remaining smaller numbers includes the acquired human TSEs such as vCJD.

In sporadic CJD the usual age of onset is late middle age (average 65 years). Most patients present with rapidly progressive dementia with focal neurological signs including ataxia, myoclonus, visual disturbances, and rigidity. Death usually occurs within 4-6 months of clinical onset.

The first case of iatrogenic transmission of CJD was identified in 1974 in a

corneal graft recipient. Since then, several hundreds of cases of iatrogenic CJD have been reported, most of which have occurred in recipients of human derived pituitary hormones or human derived dura mater grafts. Other rare sources of infection include contaminated neurosurgical instruments and intracerebral electrodes. Incubation periods for iatrogenic CJD range from 1-2 years for neurosurgical routes of transmission to over 30 years in some pituitary hormone recipients.

In 1996, the national CJD surveillance Unit in the UK identified a new form of CJD, which is known as variant CJD (vCJD). Variant CJD generally affects young adults (mean age at onset 28 years) with clinical illness that lasts on average 14 months. The initial features include psychiatric abnormalities and sensory abnormalities, which are usually followed by ataxia, myoclonus and other movement disorders and accompanied by dementia.

## 6.2. Distribution of Infectivity

In patients with sporadic CJD, the abnormal prion protein is only found in the central nervous system.

In patients with vCJD the abnormal prion protein is found in various lymphoid tissues including tonsils, appendix, spleen, and lymph nodes.

There are few cases where vCJD in recipients have followed transfusion of red cells from donors who have subsequently developed the disease and infection of a haemophiliac by a pooled plasma product. Blood and blood products should be assumed to be potentially infective though with a low incidence of risk.

## 6.3. Notification

All cases where CJD of any type is a possible diagnosis should be reported to the National CJD Surveillance Unit, the national Prion Clinic in London, and the local Consultant in Communicable Disease Control (CCDC) so that any necessary advice and action can be followed.

### **Contact address:**

National CJD Surveillance Unit

Western General Hospital

Crewe Road

Edinburgh

EH4 2XUT

Telephone: 0131 537 2128

## **National Prion Unit**

London

0203 448 4046

### **Consultant in Communicable Disease Control**

08442253557

## **6.4. Spread of TFE's and Prevention of Infection**

Available epidemiological evidence indicates that normal, social, or routine clinical contact with a patient suffering from any type of CJD, including vCJD, does not present a risk to other patients, healthcare workers, relatives, and the community.

Although cases of CJD/vCJD have been reported in healthcare workers, there have been no confirmed cases linked to occupational exposure. The highest potential risk in the context of occupational exposure is from exposure to highly infective tissues through direct inoculation (e.g.: as a result of a sharps injuries, puncture wounds or contamination off broken skin) and exposure of the mucous membranes should also be avoided.

Compliance with standard infection prevention and control precautions will minimise risks from occupational exposure.

Though the risk remains low, where correction can be achieved by other means transfusion of blood and blood products should be avoided.

Healthcare workers, who work with symptomatic patients with definite, probable, or possible CJD or vCJD or with potentially infected tissues, should be appropriately informed about the nature of the risk and relevant safety procedures.

The disease often presents after a long incubation period, where the individual is often asymptomatic but potentially infectious. Until further information is available, it is difficult to predict the number of humans who may be incubating vCJD. Thus, adequate decontamination of all instruments used in all invasive procedures is essential.

Decontamination of instruments used in invasive procedures cannot be achieved by autoclaving as this does not inactivate TSE agents. All surgical instruments that come into contact with high-risk tissues during an interventional procedure must be kept moist and separated from other instruments until they are cleaned, and then disinfected and sterilised (decontaminated). This improves the efficacy of the decontamination process and is highly cost effective. (Interventional procedures guidance [IPG666] Published date: 22 January 2020)

Thorough cleaning and physical removal of organic matter from instruments is essential. Washer disinfectors must conform to and be validated against HTM 01 - 01.

Special procedures must be followed for instruments that have or may have

been used on patients with confirmed or suspected TSE, or who are in an at-risk category (tables 2 and 3 and appendices)

Lumbar puncture kits must be single use only.

## 6.5. Identification of Patients with TSE's

It is the responsibility of the clinician to ensure that an assessment to determine risk is undertaken. Appendix 5 must be completed by patient's consultant prior to any surgical or endoscopic procedures or any invasive procedure that involves equipment that is re-processed. It is acknowledged that in an emergency situation, it may not be possible to complete the risk assessment prior to the procedure; however, this must be completed immediately after the procedure and **instruments quarantined until the assessment is completed.**

**Table 1 - Categorisation of patients by risk**

Information Category	Details of Categorisation of patients by risk
<b>Symptomatic patients</b>	<ul style="list-style-type: none"> <li>• Patients who fulfil the diagnostic criteria for definite, probable or possible CJD or vCJD.</li> <li>• 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.</li> </ul>
<b>Patients 'at increased risk' from genetic forms of CJD.</b>	<ul style="list-style-type: none"> <li>• Individuals who have been shown by specific genetic testing to be at significant risk of developing CJD.</li> <li>• Individuals who have a blood relative known to have a genetic mutation indicative of genetic CJD.</li> <li>• Individuals who have or have had two or more blood relatives affected by CJD or other prion disease.</li> </ul>
<b>Patients identified as 'at increased risk' of vCJD through receipt of blood from a donor who later developed vCJD</b>	<ul style="list-style-type: none"> <li>• Individuals who have received labile blood components (whole blood, red cells, white cells or platelets) from a donor who later went on to develop vCJD.</li> </ul>
<b>Patients identified as 'at increased risk' of CJD/vCJD through iatrogenic exposure</b>	<ul style="list-style-type: none"> <li>• Recipients of hormone derived from human pituitary glands, e.g. growth hormone, gonadotrophin, are at increased risk of transmission 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, use of human derived products may have continued in other countries after these dates.</li> <li>•</li> </ul>

Information Category	Details of Categorisation of patients by risk
	<ul style="list-style-type: none"> <li>• Individuals who underwent intradural brain or spinal surgery before August 1992 who received (or might have received) a graft of human derived dura mater are “at increased risk” unless evidence can be provided that human derived dura mater was not used.</li> <li>• 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.</li> <li>• Individuals who have received an organ or tissue from a donor infected with CJD/vCJD or “at increased risk” of CJD/vCJD.</li> <li>• Individuals identified as having received blood or blood components from 300 or more donors since January 1990.</li> <li>• Individuals who have given blood to someone who went on to develop vCJD.</li> <li>• Individuals who have received blood from someone who has also given blood to a patient who went on to develop vCJD.</li> <li>• Individuals who have been treated with certain implicated UK sourced plasma products between 1990 and 2001.</li> </ul>

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

## 6.6. Clinical Management of patients with or at increased risk of CJD/vCJD (Ward Areas)

The admission of any symptomatic or at-risk patient of having CJD/vCJD (see Table 1) must be reported to the Infection Prevention & Control Team.

There is no evidence that normal social or routine clinical contact of a CJD/vCJD patient presents a risk to healthcare workers, relatives, and others. Isolation of the patient is unnecessary, and they can be nursed on the open ward using standard infection, prevention & control precautions.

The following additional precautions must be adhered to for procedures that carry a risk of contamination with CSF or blood, for example, lumbar puncture and biopsies:

- Disposable plastic apron and gloves.
- Goggles/visor if splashing is likely to occur.

- Disposable drapes (to catch and blood spillage).
- Single use only instruments and other equipment, for example lumbar puncture kit.

The procedure should be undertaken in a treatment room by trained personnel who are aware of the hazards involved, rather than at the patient's bedside.

All clinical waste must be placed in a yellow clinical waste bag and incinerated.

Blood, biopsy, and lumbar punctures samples must be identified as a 'high risk' specimen when sending to the laboratory.

## **6.7. Clinical Management of patients with or at increased risk of CJD/vCJD (Theatres)**

It is the responsibility of the patient's Consultant to undertake a patient assessment and determine the patients risk group.

The operative procedure should only be undertaken if there is no reasonable alternative. This is especially important where flexible endoscopes are involved, as there is no way of re-processing such equipment. However, patients must not be denied treatment on the basis of their CJD/vCJD risk.

All staff involved in the care of the patient must be aware of the specific precautions and have received adequate training.

The principles for reducing the risks from percutaneous exposure to blood borne viruses apply equally to CJD.

A tracking system should be in place that will trace instruments back to all patients who have undergone a surgical procedure, including flexible endoscopes and associated equipment.

For all symptomatic patients (i.e., those who fulfil the criteria for definite, probable, or possible CJD or vCJD) the following precautions should be taken:

- The patient should be placed at the end of the list where possible.
- Inform the Infection Prevention and Control team, theatre co-ordinator, Sterile Services Manager and appropriate laboratories.
- Keep traffic in theatre to a minimum.
- Keep equipment to a minimum.
- The following single use protective clothing should be worn:
  - Liquid repellent operation gown, over a plastic apron.
  - Sterile surgical gloves.
  - Mask.

- Goggles or visor.

Use single-use disposable instruments and equipment where possible. If single use items are not available, the instruments and equipment should NOT be re used under any circumstances.

Identify specimens as ‘high risk.’

Standard infection control precautions should be followed for any spillages which should be cleared up using disposable items as quickly as possible, keeping contamination to a minimum. Disposable gloves and apron should be worn when removing such spillages. All surfaces contaminated with high-risk material e.g.: blood, CSF, or brain tissue, should be cleaned with sodium hypochlorite 10,000 ppm using standard disinfection processes.

Destroy all used instruments and protective clothing by incineration.

For asymptomatic patients at risk from familial or iatrogenic CJD, the same precautions apply.

## 6.8. Handling of Instruments that are not designated as single use.

Where single-use instruments are not available, the handling of re-usable instruments depends on a combination of the risk status of the patient, the tissue(s) involved in the procedure and the type of CJD.

The following tables 2 and 3 describe the tissue infectivity and the actions to be taken for CJD and vCJD. These actions are also summarised in the algorithms in appendix 1 and 2.

**Table 2 - Handling of Instruments for patients with or at increased risk of CJD other than vCJD**

Tissue Infectivity	Status of Patient		
	Definite or Probable	Possible	At Increased Risk
<b>High</b> Brain. Spinal cord. Cranial nerves, specifically the entire optic nerve and the intracranial components of the other cranial nerves. Cranial ganglia. Posterior eye, specifically the posterior hyaloid face, retina, retinal pigment	Single use OR Destroy by incineration OR Quarantine for re-use exclusively on the same patient	Single use OR Quarantine for re-use exclusively on the same patient pending diagnosis	Single use OR Destroy by incineration OR Quarantine for re-use exclusively on the same patient

Tissue Infectivity	Status of Patient		
	Definite or Probable	Possible	At Increased Risk
epithelium, choroid, sub retinal fluid and optic nerve. Pituitary gland.			
<b>Medium</b> Spinal ganglia. Olfactory epithelium	Single use OR Destroy by incineration OR Quarantine for re-use exclusively on the same patient	Single use OR Quarantine for re-use exclusively on the same patient pending diagnosis	Single use OR Destroy by incineration OR Quarantine for re-use exclusively on the same patient
<b>Low</b>	No special precautions	No special precautions	No special precautions

**Table 3 - Handling of Instruments patients with or increased risk of vCJD**

Tissue Infectivity	Status of Patient		
	Definite or Probable	Possible	At Increased Risk
<b>High</b> Brain. Spinal cord. Cranial nerves, specifically the entire optic nerve and the intracranial components of the other cranial nerves. Cranial ganglia. Posterior eye, specifically the posterior hyaloid face, retina, retinal pigment epithelium, choroid, sub retinal fluid and optic nerve. Pituitary gland.	Single use OR Destroy by incineration. OR Quarantine for re-use exclusively on the same patient	Single use OR Quarantine for re-use exclusively on the same patient pending diagnosis	Single use OR Destroy by incineration. OR Quarantine for re-use exclusively on the same patient

Tissue Infectivity	Status of Patient		
	Definite or Probable	Possible	At Increased Risk
<b>Medium</b> Spinal ganglia. Olfactory epithelium. Tonsil. Appendix. Spleen. Thymus. Adrenal gland. Lymph nodes and gut associated lymphoid tissues.	Single use OR Destroy by incineration OR Quarantine for re-use exclusively on the same patient	Single use OR Quarantine for re-use exclusively on the same patient pending diagnosis	Single use OR Destroy by incineration OR Quarantine for re-use exclusively on the same patient
<b>Low</b>	No special precautions	No special precautions	No special precautions

## 6.9. Quarantining of Surgical Instruments

The guidelines for quarantining of surgical instruments are kept by the theatre manager. This process should be carried out following consultation with the Sterile Services Manager.

## 6.10. Clinical Management of patient with or at increased risk of CJD (Endoscopy)

There is currently no evidence that TSEs have been transmitted through Endoscopy.

Flexible endoscopes are expensive pieces of equipment, which cannot be completely decontaminated with current methods. The risks are greater when biopsies are taken which may be contaminated with lymphoid tissue.

Single use disposable biopsy forceps should be used routinely in all patients.

The biopsy port rubber cap and channel cleaning brushes must be disposed of between patients who are symptomatic or at risk of CJD/vCJD.

A system should be in place to track endoscopes with their accessories back to individual patients.

Taking a biopsy is considered sufficient risk to quarantine the endoscope after use. Advice is therefore not to take a biopsy unless absolutely necessary.

A list of Endoscopic procedures classified as invasive or non-invasive is available on the Gov. UK website in annex F:

A summary of the precautions for flexible endoscopes is located in table 4 and 5 below. Rigid endoscopes should go to Sterile Services for autoclaving.

Endoscopes, other than those used in the CNS and nasal cavity, which have been used for invasive procedures in most individuals designated as “at increased risk” of vCJD can be returned to use after decontamination.

The endoscope should be put through all the normal stages of cleaning and be disinfected separately from other equipment within an EWD.

When decontaminating endoscope cleaning equipment, the EWD should be put through an “empty” self-disinfection cycle as per recommended routine. Provided that the cleaning equipment is decontaminated as indicated, there is no known risk of transmission of TSE agents via this route.

Following use in patients at risk of vCJD endoscopic accessories (including normally reusable devices such as heater probes) and cleaning aids such as brushes should be disposed of as healthcare waste.

Prion proteins are extremely hydrophobic, making them far more difficult to remove from instrument surfaces once they have dried on to a surface. Full endoscope decontamination should therefore commence within three hours of use. If there is a delay of more than three hours, it should be assured that the mechanism for keeping the endoscope moist until full decontamination will continue to be effective during this period.

#### 6.10.1. Summary of Precautions Advised for the use of flexible Endoscopes.

**Table 4 - CJD other than vCJD**

Tissue Infectivity	Status of Patient		
	Symptomatic		Asymptomatic
	Definite/Probable	Possible / Diagnosis Unclear	At-risk Iatrogenic / Familial
<b>High</b> Brain Spinal cord	Single use OR Destroy after use	Single use OR Quarantine pending diagnosis	Single use OR Destroy after use OR Quarantine for re-use exclusively on the same patient.
<b>Medium</b> Olfactory epithelium	Single use OR Destroy after use	Single use OR Quarantine pending diagnosis	Single use OR Destroy after use OR

Tissue Infectivity	Status of Patient		
	Symptomatic		Asymptomatic
	Definite/Probable	Possible / Diagnosis Unclear	At-risk Iatrogenic / Familial
			Quarantine for re-use exclusively on the same patient.
<b>Low/None Detectable</b> All other tissues	No special precautions**	No special precautions**	No special precautions

\* 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.

\*\* The decontamination procedures advised in this guidance, taken together with the **CFPP 01-06, NICE Interventional procedure guidance [IPG666]** and **BSG Guidelines for Decontamination of Equipment for Gastrointestinal Endoscopy** (2020), should be followed.

**Table 5 - vCJD and CJD type uncertain.**

Tissue Infectivity	Status of Patient			
	Symptomatic		Asymptomatic	
	Definite/ Probable	Possible vCJD, possible sCJD or diagnosis unclear <sup>1</sup>	At risk (blood <sup>***</sup> recipient from a donor who later developed vCJD)	At risk <sup>2</sup> Other iatrogenic
<b>High</b> Brain Spinal cord	Single use OR Destroy after use OR Quarantine <sup>3</sup> for re-use exclusively on same patient	Single use OR Quarantine pending diagnosis	Single use OR Destroy after use OR Quarantine <sup>3</sup> for re- use exclusively on same patient	Single use OR Destroy after use OR Quarantine <sup>3</sup> for re- use exclusively on same patient
<b>Medium</b> Olfactory epithelium*	Single use OR Remove from	Single use OR Quarantine	Single use OR	No special precautions unless contaminat

Tissue Infectivity	Status of Patient			
	Symptomatic		Asymptomatic	
	Definite/ Probable	Possible vCJD, possible sCJD or diagnosis unclear <sup>1</sup>	At risk (blood <sup>***</sup> recipient from a donor who later developed vCJD)	At risk <sup>2</sup> Other iatrogenic
	use OR Quarantine <sup>3</sup> for re-use exclusively on same patient	pending diagnosis	Destroy after use OR Quarantine <sup>3</sup> for re- use exclusively on same patient	ed with olfactory epithelium* If contaminat ed: single use OR destroy after use OR Quarantine <sup>3</sup> for re-use exclusively on the same index patient.
<b>Medium</b> Lymphoid tissue**	Single use OR Remove from use OR Quarantine <sup>3</sup> for re-use exclusively on same patient	Single use OR Quarantine pending diagnosis	Single use OR Destroy after use OR Quarantine <sup>3</sup> for re- use exclusively on same patient	No special precaution s <sup>□</sup>
<b>Low / None Detectable</b> All other tissues	No special precautions <sup>4</sup>	No special precautions <sup>4</sup>	No special precautions <sup>4</sup>	No special precaution s <sup>4</sup>

\* 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.

\*\*For the purposes of this Annex, 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.

- 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 of this guidance).
- 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 this guidance.
- 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 BSG Guidance for Decontamination of Equipment for Gastrointestinal Endoscopy (2020).
- The decontamination procedures advised in this guidance, taken together with the HTM01-06 or equivalent national guidance and BSG Guidance for Decontamination of Equipment for Gastrointestinal Endoscopy (2020) should be followed (<https://www.bsg.org.uk/wp-content/uploads/2021/02/BSG-Decontamination-guidance-2020-update.pdf>)

### 6.11. Clinical Management of patients with or at increased risk of CJD (Ophthalmology)

There has been one definite case of CJD transmission from corneal transplantation reported, in the US in 1974. The patient, a 55-year-old woman, received a cornea from a donor with biopsy proven sCJD.

She developed a neurologic illness eighteen months after surgery and died 8 months later. The recipient's autopsy was positive for CJD. A further probable case of CJD transmission reported in Germany in 1997 was that of a 45-year-old woman who developed clinical symptoms and EEG evidence of sCJD 30 years after keratoplasty. The donor had biopsy proven CJD, but an autopsy of the recipient was refused. Several further possible cases of CJD transmission from corneal transplantation have been reported over the past 2 decades but it is uncertain as to the significance of the corneal transplantation in their subsequent CJD disease development. There are

no other known cases of ophthalmic surgery or diagnostic procedure having resulted in CJD transmission between patients.

The risk of iatrogenic transmission of CJD/vCJD during a surgical or diagnostic procedure is dependent on the risk of tissue infectivity and the nature of the procedure itself.

**Any posterior segment eye surgery or procedure is considered high risk.**

**Any anterior segment eye surgery or procedure is considered low risk.**

The absence of any detectable abnormal prion protein in anterior segment tissue, the paucity of epidemiological evidence supporting iatrogenic transmission through anterior segment surgery and only a single definite case of iatrogenic transmission through corneal transplantation in 1974 (it is possible that the corneal tissue was contaminated by posterior segment tissue during processing) has led the ACDP TSE Working Group and its Ophthalmology Subgroup to propose anterior segment surgery or procedures as low risk for iatrogenic transmission.

There have been no known cases of iatrogenic transmission of CJD/vCJD resulting from diagnostic examination or contact lens wear. Although contact with the cornea is considered as low risk in terms of iatrogenic transmission of CJD/vCJD any steps that can further reduce potential risk of iatrogenic transmission are to be encouraged. A balance between pragmatism and a precautionary approach has been reached in the following advice.

Instruments and contact lenses considered within this section include:

- Soft refractive and therapeutic contact lenses.
- Rigid trial contact lenses, both corneal and scleral.
- Tonometer prisms (Goldmann) and other contact tonometry devices.
- Diagnostic contact lenses such as gonioscopes, fundus lenses, 3mirror lenses.
- Contact lenses used in therapy, often in conjunction with laser treatment, for example in capsulotomy, iridotomy, trabeculoplasty, retinopexy.
- A and B scan ultrasound probes.
- Electronic pachymeters.
- Electrodes used in electrodiagnostic procedures such as electroretinography.
- Prosthetic devices including trial or temporary artificial eyes.

The use of single use instruments or contact lenses is recommended for use on those designated at increased risk of CJD or vCJD. Alternatively, the instruments or contact lenses should be quarantined and used solely on that individual patient. This latter approach could be problematic in ensuring mandatory sole use and thus the single use instrument approach is considered more practical in these circumstances.

If reusable instruments or contact lenses are to be used, it is imperative that they are cleaned and decontaminated in an acceptable and consistent way. Previous guidance regarding cleaning and decontamination of lenses and tonometry prisms involving 20,000 ppm sodium hypochlorite is nationally poorly adhered to. However, it is important that it is followed, to reduce the risk of iatrogenic disease transmission.

Pragmatic solutions to aid ophthalmic practitioners in this area include:

- Having a large subset of instruments (for example Goldmann tonometry prisms) which are used once each during a clinic and following use are rinsed and kept wet. They can then be cleaned and decontaminated collectively at end of the clinic, according to the protocol in Appendix 6.
- The possibility of using wholly disposable or non-contact systems for examination.
- Encouraging procurement of non-contact or disposable covers for certain equipment, for example pachymeters or ultrasound probes.
- Discussion with the local infection control team, decontamination team and/or microbiologist should be encouraged to promote best practice.

**The practice of decontaminating tonometers with alcohol wipes alone is not sufficient to remove prion material and may in fact fix the prion protein to the surface of the instrument.**

## 6.12. Transrectal Prostatic biopsy in men at risk of vCJD

Some transrectal prostatic biopsies are undertaken by means of single use needles passed **through the internal lumens** of reusable ultrasound probes.

Patients at risk of vCJD requiring transrectal prostatic biopsy should have the procedure performed by means of single use equipment that runs alongside (rather than through) the ultrasound probe. Where a unit does not have such equipment and intends to carry out a biopsy procedure on a patient at risk of vCJD, their options are as follows:

- To refer the patient to a unit offering the alternative technique that does not pose a risk of contaminating the internal channels with traces of biopsy tissue.
- To borrow the alternative equipment from another unit.

- To undertake the procedure with equipment that has internal biopsy channels and then quarantine the reusable components of that equipment after decontamination. It must be accepted that this equipment would be unlikely to return to general use, except for dedicated re-use in the same patient.

### **6.13. Care of the Deceased**

The body should be placed in a fully sealable cadaver bag. The risk of infection should be recorded on the mortuary care record.

Mortuary Staff should be contacted via Switchboard to make them aware of the patient (if outside working hours the on-call Mortuary Technician can be contacted via switchboard).

All requests for Postmortem must be made through H M Coroner or Lead Pathologist whichever is applicable (Postmortem examination on suspected or confirmed CJD cases will not be performed at RCHT)

Embalming should be avoided in confirmed or suspect cases.

### **6.14. Recording of staff who are involved in the clinical care of CJD patients.**

Employers are required to keep a list of employees exposed to the agent of CJD of any type when there is a deliberate intention to work with the agent or, in cases of inoculation injury, if a risk assessment shows there is a significant risk. It is important to emphasise that a list is required where there is a likelihood of exposure, not simply when there has been a known incident or accident. This may include the scrub team in theatres. The lists will be kept by Occupational Health for 40 years after the last exposure. Some staff may need to be listed as being potentially exposed to CJD for example, neurosurgeons, laboratory, and mortuary staff.

A snapshot audit of compliance with the risk assessment tool will be carried out on an annual basis.

## **7. Dissemination and Implementation**

This policy will be implemented via the following routes:

- Information regarding the policy will be disseminated to the Infection Prevention and Control Link Practitioners during their quarterly updates.
- The policy will be included in the Trust's Document Library.
- The policy will be circulated to all Ward Sisters/departmental Managers and Matrons.

## 8. Monitoring compliance and effectiveness

Information Category	Detail of process and methodology for monitoring compliance
Element to be monitored	Audit of compliance with the Risk Assessment tool
Lead	Decontamination Lead
Tool	Risk assessment tool and medical records
Frequency	Annually
Reporting arrangements	Report on the audit to be submitted to the Hospital Infection Prevention and Control Committee
Acting on recommendations and Lead(s)	The Hospital Infection Prevention and Control Committee will undertake subsequent recommendations and action planning for any or all deficiencies and recommendations within reasonable timeframes.
Change in practice and lessons to be shared	Required changes to practice will be identified and actioned immediately. A lead member of the team will be identified to take each change forward where appropriate. Lessons learned will be shared with all the relevant stakeholders.

## 9. Updating and Review

This policy will be reviewed within three years.

## 10. Equality and Diversity

10.1. This document complies with the Royal Cornwall Hospitals NHS Trust service Equality and Diversity statement which can be found in the [Equality Diversity And Inclusion Policy](#) or the [Equality and Diversity website](#).

10.2. Equality Impact Assessment

The Initial Equality Impact Assessment Screening Form is at Appendix 2.

## Appendix 1. Governance Information

Information Category	Detailed Information
<b>Document Title:</b>	Management of Patients who are Symptomatic or at increased risk of transmissible Creutzfeldt – Jakob disease (CJD) Policy V2.0
<b>This document replaces (exact title of previous version):</b>	Management of Patients who are Symptomatic or at increased risk of transmissible Creutzfeldt – Jakob disease (CJD) Policy V1.0
<b>Date Issued / Approved:</b>	June 2023
<b>Date Valid From:</b>	June 2023
<b>Date Valid To:</b>	June 2026
<b>Author / Owner:</b>	Joanne Taylor, Consultant Nurse/Deputy Director of Infection Prevention and Control
<b>Contact details:</b>	01872 254969
<b>Brief summary of contents:</b>	This policy provides guidance on the management of patients who are symptomatic or are at increased risk of Creutzfeldt-Jakob disease (CJD)
<b>Suggested Keywords:</b>	Creutzfeldt – Jakob disease, Infection Prevention
<b>Target Audience:</b>	<b>RCHT:</b> Yes <b>CFT:</b> No <b>CIOB ICB:</b> No
<b>Executive Director responsible for Policy:</b>	Chief Nursing Officer
<b>Approval route for consultation and ratification:</b>	Infection Prevention and Control Committee
<b>Manager confirming approval processes:</b>	Joanne Taylor
<b>Name of Governance Lead confirming consultation and ratification:</b>	Joanne Taylor
<b>Links to key external standards:</b>	Regulation 12

<p><b>Related Documents:</b></p>	<p>Advisory Committee on Dangerous Pathogens and the Spongiform Encephalopathy (2015)</p> <p>Infection Control of CJD, vCJD and other human prion diseases in healthcare and community settings. London Department of Health,</p> <p><a href="https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf">https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf</a></p> <p>NICE (2020) Reducing the risk of transmission of Creutzfeldt–Jakob disease (CJD) from surgical instruments used for interventional procedures on high-risk tissues.</p> <p><a href="https://www.nice.org.uk/guidance/ipg666">https://www.nice.org.uk/guidance/ipg666</a></p> <p>UKHSA (2015) Patients at increased risk of CJD – background information for healthcare staff.</p> <p><a href="https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/474350/CJD_background_healthcare_staff_301015.pdf">https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/474350/CJD_background_healthcare_staff_301015.pdf</a></p> <p>British society of Gastroenterology (2013) Changed guidance on the need to quarantine endoscopes following invasive gastrointestinal endoscopy in patients at risk of vCJD. This can be found in.</p> <p><a href="#">BSG-Decontamination-guidance-2020-update.pdf</a></p> <p>Department of Health (2016) HTM 0106 Decontamination of flexible endoscopes Part A: Policy and management.</p> <p><a href="https://www.gov.uk/government/publications/management-and-decontamination-of-flexible-endoscopes">https://www.gov.uk/government/publications/management-and-decontamination-of-flexible-endoscopes</a></p> <p>Department of Health (2016) HTM 0101 Management and decontamination of surgical instruments (medical devices) used in acute care Part A: Management and provision.</p> <p><a href="https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/536144/HTM0101PartA.pdf">https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/536144/HTM0101PartA.pdf</a></p> <p>Reducing the risk of transmission of Creutzfeldt–Jakob disease (CJD) from surgical instruments used for interventional procedures on high-risk tissues.</p> <p>Interventional procedures guidance [IPG666] Published date: 22 January 2020</p> <p><a href="https://www.nice.org.uk/guidance/ipg666">https://www.nice.org.uk/guidance/ipg666</a></p>
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Information Category	Detailed Information
	Creutzfeldt-Jakob Disease (CJD) incident management guidance document and forms for trusts, hospitals and local health protection teams.  <a href="http://www.gov.uk">CJD: public health action following report of new case or person at increased risk - GOV.UK (www.gov.uk)</a>
<b>Training Need Identified:</b>	No
<b>Publication Location (refer to Policy on Policies – Approvals and Ratification):</b>	Internet and Intranet
<b>Document Library Folder/Sub Folder:</b>	Clinical / Infection Prevention and Control

### Version Control Table

Date	Version Number	Summary of Changes	Changes Made by
July 2009	1.0	New Policy	IPAC team
September 2010	2.0	Complete review and consultation. Change to title. Changes made in accordance with updated national guidance. Section on ophthalmology included.	Louise Dickinson Consultant Nurse Joint DIPC
December 2013	3.0	Changes made to endoscopy section to reflect changes in National Guidance.  Changes made to categorisation of patients by risk to reflect National Guidance.	Louise Dickinson Consultant Nurse Joint DIPC
March 2017	4.0	References updated. Changes made to endoscopy section. Section relating to transrectal prostatic biopsy added.	Louise Dickinson Consultant Nurse Joint DIPC
August 2020	5.0	Reformatted.	Jean James, IPAC Lead Nurse
November 2020	V1.0	Merge CFT/RCHT Policies	Jean James, IPAC Lead Nurse and Belinda Caslake, IPAC Team Lead
April 2023	V2.0	Full review, no change to content. Converted back to RCHT only Policy and updated on current RCHT policy template  Updated external hyperlinks for government, NICE and BSG guidance.	Gladys Marquez, IPAC Specialist Practitioner  Rashima Hamdan, IPAC Senior

Date	Version Number	Summary of Changes	Changes Made by
			Specialist Practitioner

**All or part of this document can be released under the Freedom of Information Act 2000**

**All Policies, Strategies and Operating Procedures, including Business Plans, are to be kept for the lifetime of the organisation plus 6 years.**

**This document is only valid on the day of printing.**

### **Controlled Document**

This document has been created following the Royal Cornwall Hospitals NHS Trust [The Policy on Policies \(Development and Management of Knowledge Procedural and Web Documents Policy\)](#). It should not be altered in any way without the express permission of the author or their Line Manager.

## Appendix 2. Equality Impact Assessment

### Section 1: Equality Impact Assessment (EIA) Form

The EIA process allows the Trust to identify where a policy or service may have a negative impact on an individual or particular group of people.

For guidance, please refer to the Equality Impact Assessment Policy (available from the document library) or contact the Equality, Diversity, and Inclusion Team  
[rcht.inclusion@nhs.net](mailto:rcht.inclusion@nhs.net)

Information Category	Detailed Information
<b>Name of the strategy / policy / proposal / service function to be assessed:</b>	Management of Patients who are Symptomatic or at increased risk of transmissible Creutzfeldt – Jakob disease (CJD) Policy V2.0
<b>Department and Service Area:</b>	Infection Prevention and Control
<b>Is this a new or existing document?</b>	Existing
<b>Name of individual completing EIA</b> (Should be completed by an individual with a good understanding of the Service/Policy):	Joanne Taylor
<b>Contact details:</b>	01872 254969

Information Category	Detailed Information
<b>1. Policy Aim - Who is the Policy aimed at?</b>  (The Policy is the Strategy, Policy, Proposal or Service Change to be assessed)	To provide staff with the necessary information and knowledge to effectively reduce the risk
<b>2. Policy Objectives</b>	Provide written information about transmission and prevention of TSE; Provide guidelines to risk assess suspected/ confirmed cases of vCJD/CJD; Provide guideline of safe handling and management of instruments.
<b>3. Policy Intended Outcomes</b>	Identify patients at high risk of vCJD / CJD
<b>4. How will you measure each outcome?</b>	Prevent reusable instrument used on confirmed/high risk cases of vCJD/CJD being re-used
<b>5. Who is intended to benefit from the policy?</b>	Audit of practice.

Information Category	Detailed Information
<b>6a. Who did you consult with?</b> (Please select Yes or No for each category)	<ul style="list-style-type: none"> <li>• Workforce: Yes</li> <li>• Patients/ visitors: No</li> <li>• Local groups/ system partners: Yes</li> <li>• External organisations: No</li> <li>• Other: No</li> </ul>
<b>6b. Please list the individuals/groups who have been consulted about this policy.</b>	<b>Please record specific names of individuals/ groups:</b> Infection Prevention and Control Committee
<b>6c. What was the outcome of the consultation?</b>	Policy approved
<b>6d. Have you used any of the following to assist your assessment?</b>	<b>National or local statistics, audits, activity reports, process maps, complaints, staff, or patient surveys:</b> No

## 7. The Impact

Following consultation with key groups, has a negative impact been identified for any protected characteristic? Please note that a rationale is required for each one.

Where a negative impact is identified without rationale, the key groups will need to be consulted again.

Protected Characteristic	(Yes or No)	Rationale
<b>Age</b>	No	Infections may affect any age
<b>Sex</b> (male or female)	No	Infections may affect any gender
<b>Gender reassignment</b> (Transgender, non-binary, gender fluid etc.)	No	Infections may affect any gender
<b>Race</b>	No	Infections may affect any groups.
<b>Disability</b> (e.g. physical or cognitive impairment, mental health, long term conditions etc.)	No	Infections may affect all regardless of disability
<b>Religion or belief</b>	No	Infections may affect any religion
<b>Marriage and civil partnership</b>	No	Infections may affect all people – married or otherwise

Protected Characteristic	(Yes or No)	Rationale
<b>Pregnancy and maternity</b>	No	Infections may affect any pregnant woman. Pregnant members of staff may need to take additional precautions depending on the organism involved.
<b>Sexual orientation</b> (e.g. gay, straight, bisexual, lesbian etc.)	No	Infections may affect any sexual orientation

**A robust rationale must be in place for all protected characteristics. If a negative impact has been identified, please complete section 2. If no negative impact has been identified and if this is not a major service change, you can end the assessment here.**

I am confident that section 2 of this EIA does not need completing as there are no highlighted risks of negative impact occurring because of this policy.

Name of person confirming result of initial impact assessment: Gladys Marquez

**If a negative impact has been identified above OR this is a major service change, you will need to complete section 2 of the EIA form available here:**  
[Section 2. Full Equality Analysis](#)

## Appendix 3: Diagnostic Criteria (DH, 2008)

### Diagnostic criteria

Table a of this guidance, categorises CJD patients in descending order of risk, distinguishing between **symptomatic** and **asymptomatic** patients. **Symptomatic** patients are those who fulfil the internationally accepted diagnostic criteria, set out below, for **definite, probable** and **possible** CJD or vCJD (<http://www.cjd.ed.ac.uk/criteria.htm>).

### Classification Criteria

#### Sporadic CJD

Neuropathological/immunocytochemical confirmation is required for a diagnosis of definite sporadic CJD.

**Probable** sporadic CJD patients will have rapidly progressive dementia, **and at least two** of the following four symptoms:

- Myoclonus.
- Visual or cerebellar problems.
- Pyramidal or extrapyramidal features.
- Akinetic mutism.

**plus** typical electroencephalogram (EEG) with generalised triphasic periodic complexes at approximately 1 per second

**or** clinical criteria for *possible* sporadic CJD **and** a positive assay for 14-3-3 protein in the cerebrospinal fluid (CSF).

**Possible** sporadic CJD patients will have rapidly progressive dementia, **two** of the symptoms listed in paragraph above **and** a duration of less than 2 years.

#### Iatrogenic (accidentally transmitted) CJD

**Definite** iatrogenic CJD requires a neuropathological diagnosis of CJD in a patient with a recognised risk factor for iatrogenic CJD (see box 1)

**Probable** iatrogenic CJD is defined as either a progressive predominantly cerebellar syndrome in a human pituitary growth hormone recipient, **or** a clinical diagnosis of probable CJD (see definition in paragraph B5 above) in a patient with a recognised risk factor for iatrogenic CJD (see Box B1)

#### Box 1

##### RELEVANT EXPOSURE RISKS FOR THE CLASSIFICATION AS IATROGENIC CJD

The relevance of any exposure to disease causation must take into account the timing of the exposure in relation to disease onset

Treatment with human pituitary growth hormone, human pituitary gonadotrophin or human dura mater graft

Corneal graft in which the corneal donor has been classified as definite or probable human prion disease

Exposure to neurosurgical instruments previously used in a case of definite or probable human prion disease

Transfusion of blood from a donor subsequently diagnosed with vCJD

This list is provisional as previously unrecognised mechanisms of human prion disease may occur

## Genetic TSE

Definite genetic TSE requires a neuropathological confirmation of TSE, **plus either** definite TSE in a first degree relative (i.e. a parent, child or sibling), **or** a pathogenic prion protein gene (PRNP) mutation (see Box B2).

Probable genetic TSE is defined as a progressive neuropsychiatric disorder **plus** either definite or probable TSE in a first degree relative, **or** a pathogenic PRNP mutation (see Box B2).

## Box 2

### **PRNP MUTATIONS ASSOCIATED WITH GSS\* NEUROPATHOLOGICAL PHENOTYPE**

P102L, P105L, A117V, G131V, F198S, D202N, Q212P, Q217R, M232T, 192 bpi

### **PRNP MUTATIONS ASSOCIATED WITH CJD NEUROPATHOLOGICAL PHENOTYPE**

D178N-129V, V180I, V180I+M232R, T183A, T188A, E196K, E200K, V203I, R208H, V210I, E211Q, M232R, 96 bpi, 120 bpi, 144 bpi, 168 bpi, 48 bp*del*

### **PRNP MUTATIONS ASSOCIATED WITH FFI\*\* NEUROPATHOLOGICAL PHENOTYPE**

D178N-129M

### **PRNP MUTATION ASSOCIATED WITH VASCULAR PrP AMYLOID**

Y145s

### **PRNP MUTATIONS ASSOCIATED WITH PROVEN BUT UNCLASSIFIED PRION DISEASE**

H187R, 216 bpi

### **PRNP MUTATIONS ASSOCIATED WITH NEURO-PSYCHIATRIC DISORDER, BUT NOT PROVEN PRION DISEASE**

I138M, G142S, Q160S, T188K, M232R, 24 bpi, 48 bpi, 48 bpi + nucleotide substitution in other octapeptides

\*GSS – Gertmann-Straussler-Scheinker disease

\*\*FFI – Fatal Familial Insomnia

### Variant CJD (vCJD)

**Definite** vCJD patients will have a progressive neuropsychiatric disorder **and** neuropathological confirmation of the disease, showing spongiform change and extensive PrPSc deposition with florid plaques throughout the cerebrum and cerebellum.

**Probable** vCJD patients can be classified under two sets of criteria:

1. They will have progressive neuropsychiatric disorder of a duration greater than 6 months, where routine investigations do not suggest an alternative diagnosis. They will also have **at least four** of the following five symptoms:
  - Early psychiatric symptoms (depression, anxiety, apathy withdrawal, delusions)
  - Persistent painful sensory symptoms (including both frank pain and/or unpleasant dysaesthesia)
  - Ataxia
  - Myoclonus or chorea or dystonia
  - Dementia

An EEG will not show the typical appearances of sporadic CJD, **or** no EEG has been performed and there is a symmetrical high signal in the posterior thalamus on a MRI brain scan (1 Zeidler et al 2000).

These patients would have had no history of potential iatrogenic exposure and no evidence of a familial form of TSE.

2. Alternatively, a **probable** vCJD patient will have had a progressive neuropsychiatric disorder for a period of longer than six months, where routine investigations do not support an alternative diagnosis, and where there is no history of potential of iatrogenic exposure or evidence of a familial form of TSE, **plus** a tonsil biopsy which is positive for PrPSc.

**Possible** vCJD patients will have progressive neuropsychiatric disorder of a duration greater than 6 months, where routine investigations do not suggest an alternative diagnosis, and there is no history of potential iatrogenic exposure or evidence of a familial form of TSE. They will **also** have **at least four** out of five of the symptoms listed above **and** an EEG that does not show the typical appearance of sporadic CJD **or** no EEG has been performed.

## **Patients who do not fulfil the criteria for possible CJD**

The NCJDSU have designated three additional categories for patients who are referred to the

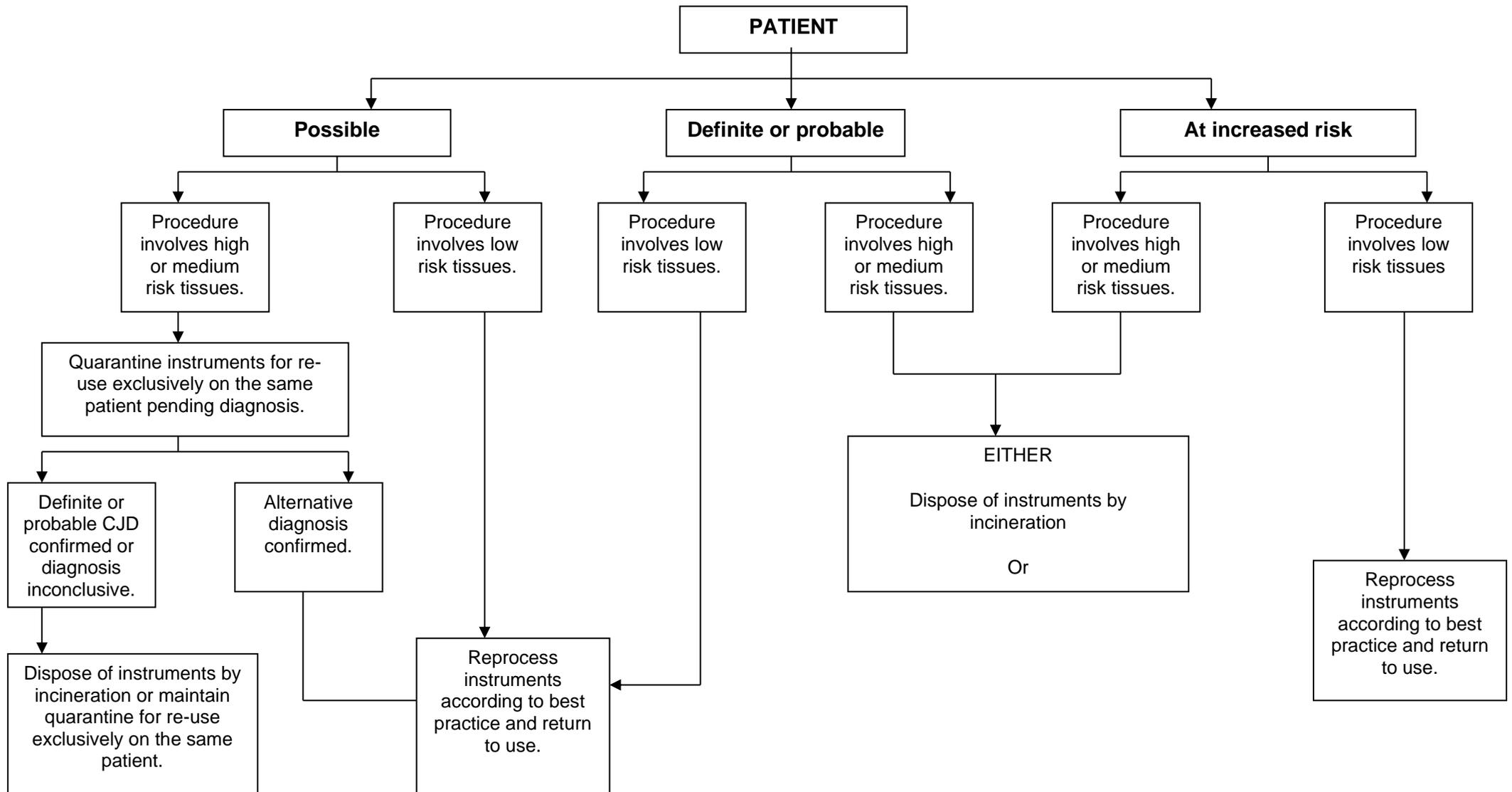
Unit but who do not meet the criteria for *possible* CJD. These can be summarised as:

**Diagnosis unclear** – the diagnostic criteria for **definite**, **probable** or **possible** CJD are not met, nor is there a reasonable alternative diagnosis. CJD therefore remains a possibility.

**CJD thought unlikely** – information indicates that a clinical diagnosis of CJD is very unlikely because of atypical disease features, and/or an atypical course, and/or atypical clinical investigation results, and/or a reasonable alternative diagnosis is made but is **not** confirmed. This category includes cases which recover clinically without a firm alternative diagnosis.

**Definitely not CJD** – information indicates that CJD is **not** the diagnosis and there is an alternative definite diagnosis proven on the basis of clinical examination, clinical investigations or pathology.

## Appendix 4. Algorithm chart for precautions for reusable instruments for surgical procedures on patients with, or “at increased risk” of, CJD, vCJD and other human prion diseases



## Appendix 5. CJD Risk Assessment – initial questions

All patients about to undergo **any** elective or emergency surgical or endoscopic procedure should be asked the question: **“Have you ever been notified that you are at increased risk of CJD or vCJD for public health purposes?”**

**Table 6: The actions to take following the patient’s response to the above question are:**

Patient’s response	Action
No	Surgery or endoscopy should proceed using normal infection control procedures unless the procedure is likely to lead to contact with high risk tissue.
Yes	Please ask the patient to explain further the reason they were notified. Special infection control precautions should be taken for all surgery or endoscopy involving contact with medium or high infectivity tissues and the infection prevention and control team should be consulted for advice. This Policy provides advice on the precautions to be taken during the treatment of patients with or at increased risk of CJD or vCJD and provides information on endoscopic procedures. The patient’s response should be recorded in their medical notes for future reference.
Unable to respond	Surgery or endoscopy should proceed using normal infection control procedures <b>unless</b> the procedure is likely to lead to contact with high risk tissue. If this is the case, please refer to the additional recommendations for high risk procedures .

**Additional recommendations for surgery which may involve contact with high risk tissue only**

**N.B. These additional recommendations are only applicable to those assessing patients in ophthalmic surgical departments for posterior ophthalmic surgical procedures.**

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

As well as asking all patients whether they have been notified as being at increased risk of CJD/vCJD, clinicians assessing patients for procedures that involve contact with high risk tissues should ask supplementary questions to assess further their CJD/vCJD risk. If a patient has answered ‘yes’ to the question in table 6 there is no additional need to ask the questions in Table 7 – the patient’s risk status has been established.

Tissues assumed or proven to have high level infectivity for CJD or vCJD are:

- Brain
- Spinal cord

- Implanted dura mater grafts prior to 1992
- Cranial nerves, specifically:
  - the entire optic nerve
  - only the intracranial components of the other cranial nerves
- Cranial nerve ganglia
- Posterior eye, specifically:
  - posterior hyaloid face
  - retina
  - retinal pigment epithelium
  - choroid
  - subretinal fluid o optic nerve
- Pituitary gland

**Table 7: CJD risk questions for patients about to undergo elective or emergency surgical procedures likely to involve contact with tissues of potentially high level infectivity**

Question to Patient		Notes to clinician
<b>1</b>	Have you a history of CJD or other prion disease in your family? If yes, please specify.	<p>Patients should be considered to be at risk from genetic forms of CJD if they have or have had:</p> <p>i) Genetic testing, which has indicated that they are at significant risk of developing CJD or other prion disease.</p> <p>ii) A blood relative known to have a genetic mutation indicative of genetic CJD or other prion disease.</p> <p>iii) 2 or more blood relatives affected by CJD or other prion disease</p>
<b>2</b>	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 iii) whether the treatment was received in the UK or in another country	<p>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 <b>discontinued in 1985</b> but human-derived products may have continued to be used in other countries. In the UK, the use of human-derived gonadotrophin was <b>discontinued in 1973</b> but may have continued in other countries after this time.</p>

Question to Patient		Notes to clinician
3	Have you ever had surgery on your brain or spinal cord?	<p>(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).</p> <p>(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 <b>who have not previously undergone high risk procedures</b>. 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.</p>

**The actions to be taken following the patient’s response to the above questions are:**

Patient’s response	Action
<b>No to all questions</b>	Surgery or can proceed using normal infection control procedures.
<b>Yes to any of questions 1, 2 or 3</b>	<p>Further investigation into the nature of the patient’s CJD risk should be undertaken, and the patient’s CJD risk assessed. This assessment of CJD risk should be recorded in the patient’s medical notes for future reference.</p> <p>If the patient is found to be at increased risk of CJD or vCJD following investigation, or the risk status is unknown at the time of the procedure, special infection control precautions should be taken for the patient’s procedure including quarantining of instruments, and the infection control team should be consulted for advice. Section 6.6 and 6.7 of this guidance provides advice for the precautions to be taken during the treatment of patients with or at increased risk of CJD or vCJD, and 6.10 provides information on endoscopic procedures. If the patient is found to be at increased risk of CJD or vCJD they should also be referred to their GP, who will need to inform them of their increased risk of CJD or vCJD and provide them with further information and advice. This is available from UKHSA: <a href="https://www.gov.uk/government/collections/creutzfeldt-jakob-disease-cjd-guidance-data-and-analysis">https://www.gov.uk/government/collections/creutzfeldt-jakob-disease-cjd-guidance-data-and-analysis</a> 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: <a href="http://www.nationalprionclinic.org/">http://www.nationalprionclinic.org/</a> 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</p>
<b>Unable to respond</b>	See below

In the event that a patient who is about to have emergency surgery is physically or otherwise unable to answer any questions, a family member, or someone close to the patient (in the case of a child, a person with parental responsibility), should be asked the CJD risk questions as set out in table 7 prior to the surgery.

If the family member, or someone close to the patient, is not able to provide a definitive answer to the CJD risk questions, the surgery should proceed but all instruments should be quarantined following the procedure. The patient's GP should be contacted after the surgery and enquiries made as to whether the patient is at increased risk of CJD/vCJD according to the questions as set out in table 7.

## Appendix 6. Guidance for the cleaning and disinfection (decontamination) of rigid contact lenses and ophthalmic medical devices which come into contact with the outer surface of the eye

1. The lens or device should be decontaminated immediately after contact with the eye surface. It should not be allowed to dry at this stage.
2. It should be rinsed in Water for Irrigation BP for not less than 30 sec.
3. It should then be cleaned on all surfaces with a liquid soap or detergent, then rinsed in Water for Irrigation BP for a further 30 sec.
4. The lens or device should then be immersed in a freshly prepared solution of sodium hypochlorite providing 10,000ppm of available chlorine for 10 min.
5. It should then be rinsed in three changes of Water for Irrigation BP for a total of not less than 10 min.
6. The device should then be shaken to remove excess water, dried with a disposable tissue, and stored dry in a suitable container.
7. Any further measure (such as autoclaving) can then be carried out if this is necessary and if the device is designed to withstand such a process. Otherwise, it is ready for immediate reuse.
8. Other chemical agents should not be used unless the device manufacturer advises against the use of sodium hypochlorite. However, agents or procedures capable of binding proteins to surfaces (e.g. isopropyl alcohol, glutaraldehyde, autoclaving) should never be used, unless devices are first decontaminated according to the above protocol.
9. The procedure described above is suitable for the great majority of devices manufactured from PMMA, glass or nonferrous metals. Where other materials are used, the manufacturer's advice should be sought.

### Notes on steps 1-6

- If circumstances do not permit the immediate decontamination of a contact lens or device, it should be immersed in Water for Irrigation BP contained in a disposable gallipot and decontaminated as soon as possible thereafter.
- Most medical devices and instruments are decontaminated with the use of potable (drinking) water. However, this has not been recommended for contact lenses and ophthalmic instruments that come into contact with the ocular surface. This is because of the risk of contamination with *Acanthamoeba* spp. and is in line with the advice given to contact lens wearers, namely, never to rinse their lenses or lens cases in tap water, and to avoid swimming and showering while wearing their lenses. The argument that rising mains water (as opposed to water from storage tanks) should be safe is not sustainable as it has been shown that rising mains pipework can become colonised by *Acanthamoeba* spp. even a short distance from its source. Moreover, in the clinical situation most

people are unaware of the source of the water available to them at their workstations or in preparation rooms, or its distance from the mains supply. For these reasons, the use of tap water has not been recommended in this guidance.

- The type of liquid soap or detergent is not specified. Unless the manufacturer publishes guidance, the advice of the local Sterile Services Department should be sought. Household detergents such as washing up liquid, and surgical scrub solutions, should not be used.
- The sodium hypochlorite solution should be prepared immediately before the episode of decontamination. Such solutions are unstable and the concentration of available chlorine diminishes with time, especially in open containers. A recommended alternative to sodium hypochlorite solutions is NaDCC (sodium dichloroisocyanurate) which is available as tablets which are mixed just before use with a dedicated diluent or with Water for Irrigation BP. NaDCC, like sodium hypochlorite, is a source of hypochlorous acid and hence of available chlorine and is widely used in healthcare environments; for example, dilutions giving 10,000 ppm available chlorine are recommended for the decontamination of blood spillages. The solution should be placed in a disposable plastic gallipot or similar disposable container. This should have a volume of not less than 50ml. (In the case of certain devices that cannot be wholly immersed, that part which comes into contact with the ocular surface must be so treated; the same consideration applies to Steps 2, 3 and 5.)
- Starch iodide paper can be used to test the final rinse water. In the presence of residual hypochlorite or chlorine, the paper that is initially white will turn purple.
- The lens or device will often have its own dedicated case for dry storage, but if not, a suitable case will have to be procured.

It is accepted that the guidance above may be difficult to adhere to in busy ophthalmic clinics. However it is important that it is followed to reduce the risk of iatrogenic disease transmission. Pragmatic solutions to aid ophthalmic practitioners in this area include:

- Having a large subset of instruments (for example Goldmann tonometry prisms) which are used once each during a clinic, and following use are rinsed and kept wet. They can then be cleaned and decontaminated collectively at end of the clinic, according to the protocol above.
- The possibility of using wholly disposable or non-contact systems for examination
- Encouraging procurement of non-contact or disposable covers for certain equipment, for example pachymeters or ultrasound probes

Discussion with the local infection control team, decontamination team and/or microbiologist should be encouraged to promote best practice

**The practice of decontaminating tonometers with alcohol wipes alone is not sufficient to remove prion material and may in fact fix the prion protein to the surface of the instrument.**