

## 31 Classical Creutzfeldt–Jakob disease

### Key points

- This chapter provides recommendations for infection control procedures to minimise the risk of transmission of classical Creutzfeldt–Jakob disease (cCJD) in health care settings.
- Variant CJD (vCJD) is excluded from the scope of this chapter as vCJD has not been reported in Australia to date. Infection control issues regarding patients with suspected or confirmed vCJD will be incorporated into Part 6, Appendix 9 once vCJD is reported in Australia and will be available on the Department of Health and Ageing website ([www.health.gov.au](http://www.health.gov.au)).
- There is presently no test available to detect cCJD infection before the onset of symptoms.
- There is no evidence that cCJD can be transmitted through normal social or sexual contact.
- The decision to implement additional precautions for equipment reprocessing is based on a risk assessment (Section 31.2.4) which incorporates the currently known infectivity of the tissue to which the instrument has been exposed (Section 31.2.2 and Table 31.1) and patient factors (Section 31.2.3 and Appendix 1 and 2). The additional precautions that may apply as a result of the risk assessment are outlined in Section 31.3 (and Table 31.2).
- Although transmission of cCJD in the health care setting is very rare, Health Care Workers (HCW) should be aware of the potential for transmission by contaminated instruments or via contaminated higher- infectivity tissues.
- The infective agent of cCJD (the prion) is resistant to routine reprocessing, making the additional precautions outlined in this chapter essential for the treatment of patients with an identified risk of cCJD infection.

### 31.1.1 Introduction

This chapter provides recommendations for infection control procedures to minimise the risk of transmission of classical Creutzfeldt–Jakob disease (cCJD) in health care settings.

The infective agent of cCJD (the prion) is resistant to routine reprocessing (as defined in AS/NZS 4187 Cleaning, disinfecting and sterilizing reusable medical and surgical instruments and equipment, and maintenance of associated environments in health care facilities). This makes the additional precautions outlined in this chapter essential for treatment of patients with an identified risk of cCJD infection.

The decision to implement additional precautions for equipment reprocessing is based on the currently known infectivity of the tissue to which the instrument has been exposed (see Table 31.1) and patient risk factors (see Appendix 1 and 2). Alternative diagnostic and management strategies, if suitable and available, should be considered in patients at risk of cCJD, provided that the care of the patient is not compromised.

Continual advances in instrument design and reprocessing technology mean that recommendations to minimise the risk of cCJD transmission in health care settings should be regularly updated. Health care establishments should ensure that they have the most current version of this chapter by checking the Department of Health and Ageing website ([www.health.gov.au](http://www.health.gov.au)).

Variant CJD (vCJD) is excluded from the scope of this document as vCJD has not yet been reported in Australia. Separate Infection Control Guidelines for vCJD address infection control issues regarding patients with suspected or confirmed vCJD and will be released on

the Department of Health and Ageing website if vCJD is reported in Australia ([www.health.gov.au](http://www.health.gov.au)) for incorporation into Part 6, Appendix 9 of the *Infection Control Guidelines for the Prevention of Transmission of Infectious Diseases in the Health Care Setting* (these guidelines). If you suspect a case of vCJD, contact your local State or Territory Health Department immediately.

### **31.1.2 Disease Categories**

For simplicity, the term ‘classical CJD’ (cCJD) is used to describe all forms of human TSE (except vCJD), including (Collins *et al* 2001, 2004, Brown *et al* 2000):

- 1) Sporadic CJD
- 2) Inherited CJD
  - a) Familial CJD
  - b) Gerstmann-Sträussler-Scheinker Disease (GSS)
  - c) Fatal Familial Insomnia (FFI)
- 3) Acquired CJD
  - a) Health care associated (iatrogenic) CJD
  - b) Kuru

### **31.1.3 Diagnosis**

There is currently no minimally invasive test available to detect cCJD infection before the onset of symptoms. There is a pre-symptomatic period during which disease transmission is presumed to be possible. Definitive diagnosis of cCJD is by neuropathological examination of brain tissue following biopsy or autopsy. However, brain biopsy is not recommended as a routine procedure to confirm the clinical suspicion of cCJD.

Methods that may assist in diagnosis of cCJD and in excluding other causes of subacute dementia in symptomatic patients include (Zerr *et al* 2000, Shiga *et al* 2004):

- electroencephalograph (EEG);
- the presence of protein 14-3-3 in cerebrospinal fluid (CSF); and
- imaging techniques such as computerised tomography (CAT Scan) and magnetic resonance imaging (MRI).

## **31.2 Assessing the risk**

The application of additional precautions to minimise the risk of transmission of cCJD is based on a risk assessment. The tissues or body fluids likely to be exposed during a procedure should be classified according to Section 31.2.2 (and Table 31.1) and the patient risk category should be identified according to Section 31.2.3. A risk assessment should then be performed according to Section 31.2.4. The additional precautions that may apply as a result of the risk assessment are outlined in Section 31.3 (and Table 31.2).

### **31.2.1 Modes of transmission**

Most cases of cCJD are sporadic. However, there is evidence of iatrogenic transmission through neurosurgical instruments contaminated with central nervous system (CNS) tissue and through contaminated tissue implants or products (dura mater grafts, corneal grafts, pituitary products). Although transmission of cCJD in the health care setting is very rare, HCW should be aware of the potential for transmission from patient to patient by contaminated instruments or via contaminated tissues. There is no epidemiological evidence to indicate that HCW are at an increased occupational risk for cCJD. There is no epidemiological evidence that cCJD can be transmitted through normal social or sexual contact, mother-to-child transmission or via blood or blood products (Brown *et al* 1994, Collins *et al* 1999, Tamai *et al* 1992, Gajdusek 1977, Will 1993, Wientjens *et al* 1996).

### **31.2.2 Infectivity of human tissues**

Table 31.1 is a guide to the known or predicted infectivity of body tissues and fluids of symptomatic and asymptomatic patients with cCJD. This information is based largely on studies of experimentally transmitted cCJD in non-human primates and other animals. Whilst there is likely a spectrum of infectivity from very low to medium to high infectivity, the classifications in Table 31.1 group the tissues and fluids according to the reprocessing that will be required after contact with these tissues (Brown 1994).

**Table 31.1 Known or predicted infectivity of human body tissues and fluids for cCJD**

Infectivity category	Tissues	Secretions and excretions
<b>High-infectivity or medium-infectivity</b> <sup>(1)</sup> (Higher-infectivity)	Brain Dura mater Pituitary gland Spinal cord Posterior eye (including retina, vitreous humour and optic nerve) Cranial and dorsal root ganglia Olfactory epithelium	
<b>Lower-infectivity or no detectable infectivity</b> <sup>(2)</sup> (Lower-infectivity)	Cornea <sup>(3)</sup> Anterior chamber of eye <sup>(3)</sup> Kidney Liver Lung Lymph nodes/spleen Placenta Uterus Adipose tissue Adrenal gland Blood & blood products Bone marrow Oral tissue (teeth, gingival tissue, dental pulp) Heart muscle Intestine Peripheral nerve Prostate Skeletal muscle Testes Thyroid gland	CSF Amniotic fluid Faeces Breast milk Nasal mucus Saliva Semen Serous exudate Sweat Tears Urine

Source: Modified from WHO *Guidelines on Tissue Infectivity Distribution in Transmissible Spongiform Encephalopathies* (2006) and the UK *Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection* (2003).

<sup>(1)</sup> Referred to in this document as ‘Higher-infectivity’ tissues. Considerable Risk of Transmission (instruments having contact with these tissues will require additional reprocessing precautions- See Appendix 4).

<sup>(2)</sup> Low Risk of Transmission (instruments having contact with these tissues and fluids only, do not require additional reprocessing precautions- See Appendix 4).

<sup>(3)</sup> It is recommended that single use instruments be used in known high risk patients (Appendix 1, for risk assessment see Section 31.2.4).

### 31.2.3 Patient risk categories

The following risk categories identify individuals who may pose a risk of transmitting cCJD:

- **High-risk** - people who represent a *definite* risk of cCJD transmission (**Appendix 1**). These patients are generally showing neurological symptoms;
- **Low-risk** - people who represent a *potential* risk of cCJD transmission (**Appendix 2**). These patients may be showing neurological symptoms or may have an identified risk factor;

(NOTE: Individuals who have been contacted by a Health Department as part of a look-back procedure from exposure to surgical instruments that had previously been used on high or medium infectivity tissues from patients later found to have contracted cCJD are likely to have a very low, but unquantifiable risk for cCJD. Until further information on the likely risk of these individuals is available, they are conservatively placed in a low risk category.)

- **Background risk** - the general population who represent no identified increased risk of cCJD transmission.

### 31.2.4 Risk assessment

Diagnostic and therapeutic procedures are divided into those where higher-infectivity tissue is exposed and those where only lower-infectivity or no detectable infectivity tissue is exposed (see Table 31.1). Patients are divided into those with a high risk, those that are considered low risk and those with background risk.

**Additional precautions (Section 31.3) are implemented when the patient is identified as being in a high- or low-risk category AND when the diagnostic or therapeutic procedure used involves the exposure of higher-infectivity tissues.**

Patient risk category	Procedures involving exposure to higher-infectivity tissues (see Table 31.1)	Procedures involving exposure to lower or no detectable infectivity tissues
<b>High-risk patient</b>	Use additional precautions	Use routine reprocessing precautions
<b>Low-risk Patient</b>	Use additional precautions	Use routine reprocessing precautions
<b>Background risk patient</b>	Use routine reprocessing precautions	Use routine reprocessing precautions

It is recommended that all patients undergoing surgical or diagnostic procedures in which higher-infectivity tissue will be exposed (eg. neurosurgery, spinal cord surgery, ophthalmic surgery, pituitary surgery) should have their cCJD risk status (high-risk, low-risk, background-risk) determined prior to the procedure.

**A template for a questionnaire to determine cCJD risk status is included in Appendix 3.**

Questionnaires should be administered to patients by the health care practitioner conducting the procedure, prior to consent for the planned procedure, and the completed questionnaire included in the patient medical record. If, on the basis of responses to the questionnaire, the patient is determined to be in a high- or low-cCJD risk category, the planned procedure may be modified or a process initiated for the implementation of additional precautions for equipment reprocessing/disposal. Health care establishments should establish systems to ensure that risk assessment, where recommended, is undertaken and documented eg. linking the process to the health care establishment booking process.

Each health care establishment should have an action plan in place, so that if the questionnaire identifies a patient with a risk of cCJD, patient admission and treatment is not delayed. There is a need to ensure that patient care is not compromised and that any reasons for variations or delays in treatment are explained to the patient in order to encourage patients with identified risk factors to disclose their risk status to health care establishments.

**A flow chart ‘Summary of Actions for a Surgical Procedure- cCJD Risk Assessment’ is included in Appendix 4.**

### **31.3 Additional Precautions**

**Additional precautions are implemented when the patient is identified as being in a high- or low-risk category AND when the diagnostic or therapeutic procedure used involves the exposure of higher-infectivity tissues.**

Relevant additional precautions that apply to the handling and reprocessing of surgical instruments and diagnostic equipment are shown in Table 31.2. For routine hospital, long-term residential or community care not involving exposure to higher-infectivity tissues, routine reprocessing procedures are all that are required for the management of cCJD patients.

#### **31.3.1 Reasons for additional precautions**

The ‘prion’, which is the infectious agent of cCJD, is resistant to routine reprocessing. The chemicals known to have some activity against prions include anionic detergents, hypochlorites and harsh acids and alkalis. However, their practical effectiveness and use in reprocessing is influenced by prior cleaning and prion strain. Occupational health issues surrounding the use of harsh acids and alkalis and potential for damage to instruments and equipment mean they are not recommended for use in reprocessing. (Brown *et al* 1982, Fichet *et al* 2004, Gibbs *et al* 1978, Jackson *et al* 2005, Tateishi 1980, Tateishi *et al* 1988, Taylor 1987, Taylor 2000).

**Table 31.2 Additional precautions required for diagnostic or therapeutic procedures involving higher- infectivity tissues for patients in the high- and low-risk categories for cCJD**

Activity	Precautions
<b>Instrument reuse</b>	Incinerate* instruments immediately after use <b>OR</b> Reprocess reusable instruments separately and keep for the exclusive use of an individual patient involved in a course of therapy (then incinerate* when no longer required) <b>OR</b> For those low risk patients who are awaiting determination of risk status as either high risk or background risk (see Appendix 2); reprocess reusable instruments separately and quarantine instruments pending determination of risk status of patient (then incinerate* if deemed high- or low- risk, <b>or</b> reprocess and put back into circulation if risk is found to be background)
<b>Intra-operative handling of instruments</b>	Instruments used on higher- infectivity tissues should be separated from general instruments and equipment during the operative procedure, and should also be quarantined from other instruments in the reprocessing area to reduce the possibility of cross contamination.
<b>Operating room set up</b>	Cameras and other equipment not in contact with the higher- infectivity tissue should be covered in plastic to protect from splashing. Surgical drapes and plastic covers should be incinerated after use.*
<b>Personal protective equipment (PPE)</b>	All HCW should wear fluid repellent single-use PPE including gloves, gowns and full face shield if higher- infectivity tissue is exposed. PPE should be destroyed by incineration* after use.
<b>Scheduling of patients</b>	Operations or procedures should be scheduled to allow for appropriate cleaning of facilities.
<b>Collection of specimens</b>	Specimens should be collected into a secure-closing container and enclosed in a plastic bag for transportation. The container should be clearly labelled with patient identification details, including a cCJD risk alert to laboratory workers and other HCWs.
<b>Anaesthetic equipment</b>	Routine reprocessing
* or appropriate alternate approved method of medical waste destruction.	

### **31.3.2 Additional precautions for destruction of equipment by incineration**

Single use instruments should be used where possible. Contaminated articles should be placed immediately into the correct clinical waste container for disposal by incineration or alternate approved method of medical waste destruction (see Chapter 15.2). Needles, blades and other sharp articles should be placed in non-reusable sharps containers (in accordance with AS/NZS 4187:2003 *Cleaning, disinfecting and sterilizing reusable medical and surgical instruments and equipment, and maintenance of associated environments in health care facilities*) and destroyed by incineration.

### **31.3.3 Additional precautions for reprocessing procedures**

Thorough washing and cleaning with anionic detergents will reduce the level of instrument contamination by all micro-organisms and therefore would be expected also to decrease the risk of transmission of prions if any were present. High-level disinfectants such as glutaraldehyde, however, enhance the adherence of prions to surfaces, and thus are contraindicated for use on instruments that may potentially be contaminated by prions (Fichet *et al* 2004, Jackson *et al* 2005).

Instruments and equipment that need to be quarantined or kept for exclusive use a particular patient and have been exposed to higher-infectivity tissues should be reprocessed according to AS/NZS 4187 *Cleaning, disinfecting and sterilizing reusable medical and surgical instruments and equipment, and maintenance of associated environments in health care facilities* with the following additional recommendations:

- Instruments that have been in contact with higher- infectivity tissues in high- or low-risk patients should be separated from other instruments in the operating room to avoid cross-contamination;
- The cCJD prion may be stabilised by drying on metal surfaces, thereby becoming more difficult to inactivate. To prevent drying, instruments potentially contaminated with higher- infectivity tissue should be immersed in a dedicated container in sterile water until they are reprocessed (for subsequent quarantine or exclusive use in that patient);
- Instruments should be cleaned in anionic detergent prior to further reprocessing;
- Contaminated instruments should be reprocessed in a separate batch, and not mixed with other surgical instruments at any stage of the reprocessing cycle;
- Ultrasonic cleaners may be used during reprocessing;
- Steam sterilisation at 134°C for 3 minutes is recommended;
- Items that have been identified as difficult to clean should be destroyed; and
- Equipment reprocessing staff should wear gloves, fluid-repellent gowns, masks and eye protection at all times when handling higher- infectivity tissues and instruments exposed to higher- infectivity tissues. After use, personal protective equipment (PPE) should be destroyed by either incineration or an appropriate alternate approved method of medical waste destruction.

- While alternative methods of reprocessing (proteases or alkaline detergents) are being actively researched, they are not yet being recommended as alternatives to destruction for instruments used on high- or low-risk patients in higher- infectivity tissues. In the future, it is likely that if a cleaning method is found to be effective in removing prions, it may be incorporated into routine reprocessing for all surgical instruments. Health care establishments should consider this when purchasing new instrument cleaning systems.

**Important note- Ventriculosopes used on high- or low risk patients must be destroyed by incineration or kept for exclusive use in that patient. All other endoscopes may be reprocessed using routine reprocessing.**

### **31.3.4 Additional precautions for tracking of reusable instruments**

All procedures exposing higher- infectivity tissues, and companies that provide loan equipment, demonstration equipment or trial equipment for use in these procedures, should have systems in place to track individual instruments and equipment to the level of the individual patient. Tracking of instruments and trays will minimise the number of patients implicated in a look-back (Section 31.4.3), where a background risk patient is subsequently diagnosed with cCJD.

Any tracking and quarantine system must minimise the risk of accidental re-introduction of potentially infected equipment and instruments into the reprocessing area.

### **31.3.5 Additional precautions for quarantine of reusable instruments and equipment**

Quarantine of equipment is the process by which instruments are separated, reprocessed, labelled and held aside for either of two courses of action; destruction or return to circulation. Quarantine of equipment should be used if the patient's cCJD risk status is not known, including during an investigation by the State or Territory Health Department. The equipment should be quarantined until the risk status is clarified.

Once the risk status of a low risk patient is determined, equipment should be either returned to circulation after reprocessing or destroyed by incineration or alternate approved method of medical waste destruction. In some instances, the National CJD Incident Panel (see Section 31.4.2) may recommend additional reprocessing before instruments or equipment are returned to circulation. If a patient is categorised as either low-risk or high-risk for cCJD, the equipment may be quarantined for future exclusive use with that patient, and then destroyed by incineration or alternate approved method of medical waste destruction when no longer required.

### **31.3.6 Additional precautions for environmental cleaning of the operative area**

Unless a spill of higher- infectivity tissues has occurred, routine containment and cleaning procedures should be used for the whole operative area, including surfaces. A spills kit (that includes occupational health and safety recommendations) should be available in areas where higher-infectivity tissues may be exposed, such as operating rooms, mortuaries and laboratories.

Contamination by spillage of higher-infectivity tissues from patients in either the low- or high-risk cCJD categories should be cleaned by first exposing the area to freshly prepared 1M sodium hydroxide (NaOH) or 20,000ppm (free chlorine) sodium hypochlorite for 1 hour at ambient temperature, followed by a rinse with water. Where surfaces cannot tolerate NaOH or hypochlorite, cleaning using anionic detergent and water will partially reduce infectivity by dilution.

Staff should be appropriately trained in cleaning of the operative area and in use of 1M NaOH and 20,000ppm sodium hypochlorite. Material Safety Data Sheets (MSDS) should be available for 1M NaOH and 20,000ppm sodium hypochlorite.

### **31.3.7 Occupational exposure to higher- infectivity tissues**

There are no additional requirements following occupational exposure to tissues of an individual with cCJD or one in the high- or low-risk category.

## **31.4 Surveillance**

cCJD is now a notifiable disease in all States and Territories in Australia. Each State and Territory will have requirements for reporting notifiable diseases, including cCJD, and methods for providing advice regarding infection control issues. See Appendix 6 for State and Territory Health Department contact details.

### **31.4.1 Surveillance by the Australian National CJD Registry (ANCJDR)**

The Australian Government Department of Health and Ageing established the ANCJDR in 1993, based in the Department of Pathology at the University of Melbourne. The registry assists the department with the ongoing surveillance of cCJD cases in Australia, identifies cCJD risk factors for population health and should be involved in suspect cCJD cases by public health authorities. The contact details of the registry are provided in Appendix 6.

### **31.4.2 Surveillance for Adverse Event Management**

Since there is no test to reliably detect cCJD prior to the onset of symptoms, it is possible that surgical instruments used on a patient with asymptomatic cCJD might subsequently be used unknowingly on other patients after routine reprocessing, with a potential risk of transmission.

In the event of patients being exposed to instruments that have previously been exposed to higher- infectivity tissues in a patient that is subsequently found to have cCJD, the following should be immediately notified:

- the executive of the health care establishment; and
- the State or Territory Health Department (see Appendix 6).

In September 2004, the Australian Government established a National CJD Incident Panel. This panel provides expert advice in the event of an adverse event involving cCJD. The relevant State or Territory Health Department assumes responsibility and is accountable for determining action to be taken, the investigation, equipment management, patient risk assessment and the scope of a look-back investigation if it is required. The Health Department may request advice from the National CJD Incident Panel on specific look-back and infection control issues.

If equipment having direct contact with higher-infectivity tissue (Table 31.1) has been used in the past on a patient subsequently found to have cCJD, the equipment should be identified and withdrawn pending a decision from the State or Territory Health Department who may obtain advice from the National CJD Incident Panel. Upon this decision, the instruments will either be destroyed or returned to use following reprocessing. Other equipment that has not been in contact with a higher-infectivity tissue should not be withdrawn and should continue to be reprocessed using routine methods.

### **31.4.3 Look-back**

The need for a look-back is determined by a risk assessment process undertaken by the State or Territory Health Department. A flow chart summarising the essential steps in a look-back procedure is provided in Appendix 5. The State or Territory Health Department in consultation with the health care establishment is responsible for tracing individuals suspected of exposure to cCJD. The National CJD Incident Panel is available to provide expert advice to inform decisions on the need for a look-back and infection control measures.

A plan for the look-back should be developed that allows for tracing of potentially exposed individuals and assessment of their potential exposure to risk. Consideration should be given to maintenance of confidentiality of patient details and the way in which information is provided (personal phone communication, face-face, written information), standardised or individualised information and involvement of the media.

### **31.4.4 Organs and tissues for transplantation**

In all situations, the following people should be excluded from the routine donation of organs and tissues:

- people in the high-risk group (Appendix 1);
- people in the low-risk group (Appendix 2) (NB: tissues are excluded from donation, but organs may be allowed to be donated, if informed consent is given by the recipient);
- people who die in psychiatric establishments, with the exception of those in whom cCJD has been specifically excluded
- people who die of dementia; and
- people who die with any obscure undiagnosed neurological disorder.

Agencies that are responsible for recruiting organ/tissue donors and for the banking of tissues should be aware of the public health implications of cCJD and should have donor exclusion criteria and screening procedures in place, in accordance with the State or Territory transplantation legislation. The Transplantation Society of Australia and New Zealand ([www.racp.edu.au/tsanz](http://www.racp.edu.au/tsanz)) have an example organ donation referral form.

Material from patient groups at risk of transmitting cCJD should not be used for the preparation of any therapeutic product or laboratory reagent (eg. thromboplastin or Kveim test material).

## 31.5 Infection Control in other settings

### 31.5.1 Dentistry

Oro-facio-maxillary surgical procedures that come into contact with **higher-infectivity** tissues in patients of high- or low- risk should be treated with additional precautions (Table 31.2). These procedures would include: (An example of higher- infectivity tissue exposed is provided in brackets)

- Major oral surgery procedures such as a maxillectomy involving orbit enucleation (optic nerve);
- Injection of the trigeminal ganglion (potential brain tissue, central nerve exposure);
- Oral surgical cancer procedures also combining a neurosurgical approach would involve exposure to tissue of higher-infectivity (potential brain tissue, central nerve exposure).

In all patients, including high- and low-risk patients (Appendix 1 and 2), instruments in contact with lower infectivity tissues (Table 31.1) through routine dental procedures can be routinely reprocessed. Dentists should take an appropriate medical history of all patients. Dentists who have patients identified as high- or low-risk should contact their State or Territory Health Department and the Australian Dental Association for additional advice on infection control procedures. Dental work on high- or low-risk patients involving exposure to higher-infectivity tissues should be performed at an establishment with HCW who are familiar with cCJD infection control procedures.

### 31.5.2 Post Mortem Examinations

Additional precautions should be used for post mortem examinations involving exposure to high infectivity sites in patients with suspect cCJD or of high- or low risk, as per Table 31.2 (Bell and Ironside 1993, Budka *et al* 1995). A set of instruments dedicated to suspect cCJD patients should be used and kept separate to instruments used to harvest organs and tissues for donation.

Removal of the brain with either an electric bone saw or a hand saw should be performed with sufficient containment to avoid aerosol production.

All tissue samples from higher-infectivity sites should be treated as potentially infectious for cCJD until proved otherwise. Tissue or fluid samples should be collected into sealed containers with the cCJD risk status of the patient clearly labelled. High infectivity tissues should be handled under Physical Containment Level 2 (PC2). Due to the resistance to inactivation by aldehydes and alcohols, brain specimens should be fixed in 4% formaldehyde solution (10% formal saline), followed by immersion in formic acid (>96%) for one hour. For

machine processing, tissues should be washed in formalin to prevent damage to containers by formic acid. For hand processing, tissues can be transferred directly from formic acid to ascending alcohol solutions.

Following post mortem, bodies should be sealed in plastic to avoid fluid leakage. Embalming of bodies should be avoided. Cadavers from high- or low-risk patients should not be used for teaching purposes.

Mortuary facilities with staff appropriately trained in cCJD infection control procedures should be available in capital cities and major regional centres in each State and Territory. Each State and Territory should have appropriate guidelines and procedures for funding post mortems for suspect cCJD patients and appropriate guidelines and procedures for funding transport of bodies of suspect cCJD patients to and from post mortem facilities.

## Appendix 1: Individuals in the high-risk category for cCJD

Classification of cCJD	Clinical signs and risk factors
<p><b>1 Sporadic TSE</b></p> <p><b>1.1 Definite</b> Neuropathologically/ immunocytochemically confirmed</p> <p><b>1.2 Probable</b> 1.2.1 I and 2/4 of II and III 1.2.2 Possible and positive 14-3-3 CSF assay</p> <p><b>1.3 Possible</b> I and 2/4 of II and duration &lt;2 years</p>	<p><b>Clinical signs</b></p> <p>I Rapidly progressive dementia</p> <p>II A Myoclonus B Visual or cerebellar problems C Pyramidal or extrapyramidal features D Akinetic mutism</p> <p>III Typical EEG</p>
<p><b>2 Accidentally transmitted TSE</b></p> <p><b>2.1 Definite</b> Definite TSE with a recognised health care acquired risk factor</p> <p><b>2.2 Probable</b> 2.2.1 Progressive predominant cerebellar syndrome in human pituitary hormone recipients 2.2.2 Probable TSE with recognised health care acquired risk factor</p>	<p><b>Recognised health care acquired risk factors</b></p> <ul style="list-style-type: none"> <li>• Treatment with human pituitary growth hormone, human pituitary gonadotrophin or human dura mater graft.</li> <li>• Corneal graft in which the corneal donor has been classified as definitely or probably having a human prion disease.</li> <li>• Exposure to surgical instruments that have come into contact with higher-infectivity tissues previously used in a case of definite or probable human prion disease.</li> </ul> <p>The relevance of any exposure to disease causation must take into account the timing of exposure in relation to disease onset. This list is provisional, as previously unrecognised mechanisms of human prion disease may occur.</p>
<p><b>3 Genetic TSE</b></p> <p><b>3.1 Definite</b> 3.1.1 Definite TSE and definite or probable TSE in first- degree relative 3.1.2 Definite TSE with a pathogenic PRNP mutation</p> <p><b>3.2 Probable</b> 3.2.1 Progressive neuropsychiatric disorder and definite or probable TSE in first-degree relative 3.2.2 Progressive neuropsychiatric disorder and pathogenic PRNP mutation</p>	<p><b>PRNP mutations</b></p> <ul style="list-style-type: none"> <li>• PRNP mutations associated with GSS neuropathological phenotype: P102L, P105L, A117V, G131V, F198S, D202N, Q212P, Q217R, M232T, 192 bpi</li> <li>• 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 deletion</li> <li>• PRNP mutations associated with FFI neuropathological phenotype: D178N-129M</li> <li>• PRNP mutation associated with vascular PRP amyloid: Y145S</li> <li>• PRNP mutations associated with proven but unclassified prion disease: H187R, 216 bpi</li> <li>• Mutations associated with neuropsychiatric disorder but not proven prion disease: I138M, G142S, Q160S, T188K, M232R, 24 bpi, 48 bpi, 48 bpi + nucleotide substitution in other octapeptides</li> <li>• PRNP mutations without clinical and neuropathological data: T188R, P238S</li> <li>• PRNP polymorphisms with established influence on phenotype: M129V</li> <li>• PRNP polymorphisms with suggested influence on phenotype: N171S, E219K, 24 bp deletion</li> <li>• PRNP polymorphisms without established influence on phenotype: P68P, A117A, G124G, V161V, N173N, H177H, T188T, D202D, Q212Q, R228R, S230S</li> </ul>

**Other** The following people are also classified as being at high risk: carriers of disease-linked mutations of the PrP gene; and persons in whom the PrP gene has not been sequenced but who have two or more first degree relatives with cCJD (including GSS or FFI). **Note:** People who have had the PrP gene sequenced and are shown not to carry the disease-linked mutation can be classified as ‘background’ risk, unless they have other demonstrated risk factors.

**(Kovacs *et al* 2002, Kovacs *et al* 2005).**

## Appendix 2: Individuals in the low-risk category for cCJD

<p>People with a progressive neurological illness of less than one year’s duration, with or without dementia for whom a determination to assign a high-risk status or background risk status cannot be made following competent professional review.</p>
<p>People with a progressive neurological illness of less than one year’s duration, with or without dementia awaiting the outcome of a professional review to assign a high-risk status or background risk status.</p>
<p>Patients undergoing a diagnostic brain biopsy for progressive brain disease or patients undergoing neurosurgical investigations (including brain biopsy) or therapeutic procedures for a progressive disorder that includes dementia.</p>
<p>All genetically related members of any family in which there is a strong family history (two or more first-degree relatives) of dementia or neurological illness, and in which affected individuals have not been competently and completely assessed neurologically, specifically for cCJD.</p>
<p>Recipients of cadaver-derived human pituitary hormones (growth hormone and gonadotrophins) before 1986.</p>
<p>Recipients of dura mater homografts or transdural neurosurgery before 1990, or neurosurgical patients for whom the use of dura mater homografts cannot be excluded by reference to patient records.</p>
<p>Individuals who have been contacted by a Health Department as part of a look-back procedure from exposure to surgical instruments that had previously been used on high or medium infectivity tissues from patients later found to have contracted cCJD are likely to have a very low, but unquantifiable risk for cCJD that is thought to be above background risk. Until further information on the likely risk of these individuals is available, they are conservatively placed in a low risk category.</p>

## Appendix 3

### Classical Creutzfeldt-Jakob Disease (cCJD) Risk Assessment Tool

#### INTRODUCTION

The following questions should be asked of a patient prior to undergoing surgery, investigations or a procedure involving any of the following higher-infectivity tissues:

- (a) Brain, pituitary or dura mater
- (b) Cranial and dorsal root ganglia
- (c) Spinal cord
- (d) Eye (Retina/Optic Nerve)
- (e) Olfactory Epithelium

*NB: if this is a repeat procedure and the following questions have already been answered, then they need not be completed again providing the patient's neurological condition remains unchanged.*

#### MEDICAL OFFICER QUESTIONS TO DETERMINE RISK STATUS

Q1. Do you think the patient may have cCJD?  Yes  No

Q2. Has the patient had a first degree relative with cCJD?  Yes  No

Q3. Does the patient have an unexplained progressive neurological illness  
of less than 12 months?  Yes  No

Q4. Does the patient have a history of receiving human pituitary hormone for  
infertility or human growth hormone for short stature (prior to 1986)?  Yes  No

Q5. Has the patient previously had surgery on the brain or spinal cord that  
included a dura mater graft (prior to 1990)?  Yes  No

Q6. Has the patient been involved in a 'look-back' for cCJD or shown you  
a 'medical in confidence letter' regarding their risk for cCJD?  Yes  No

**Action: If the patient answers yes to any of the above questions, please contact infection control personnel in your health care establishment. Put into place the action plan for potential cCJD patients.**

I have undertaken the appropriate action as required by the health care establishment infection control policies regarding cCJD.

Name of the Health Care Practitioner

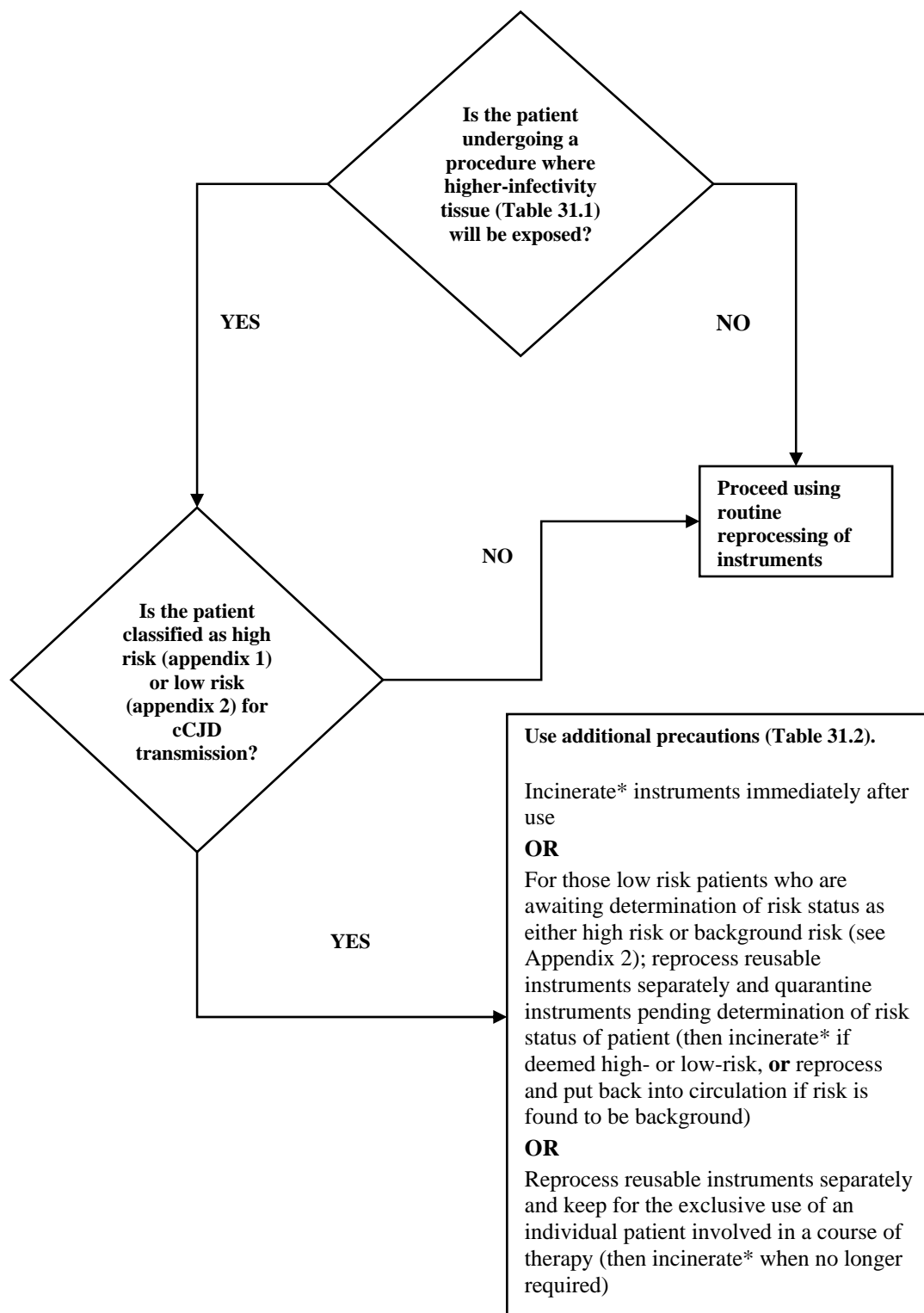
Signature

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## Appendix 4

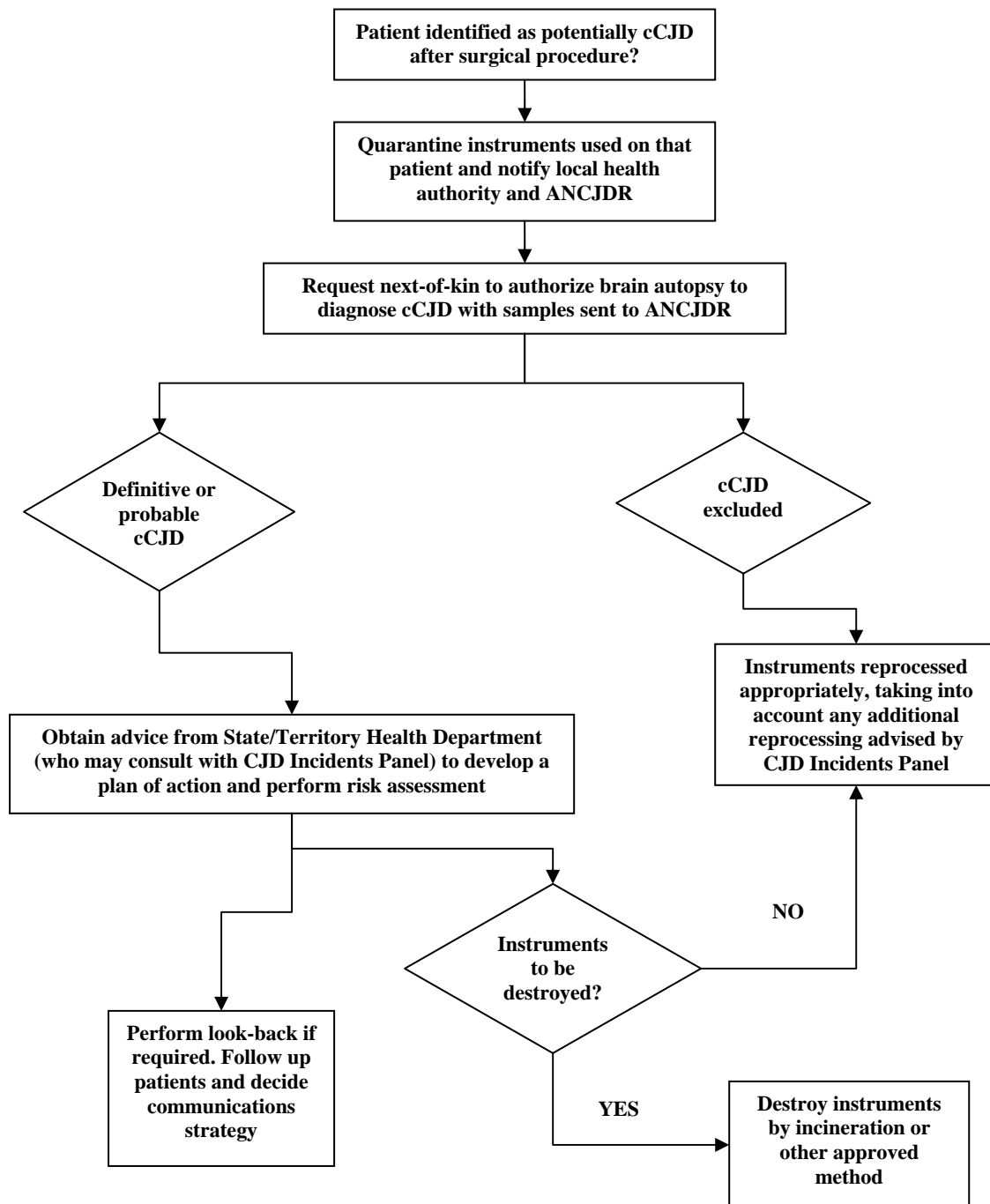
### Summary of Actions for a Surgical Procedure – cCJD Risk Assessment



\* or appropriate alternate approved method of medical waste destruction.

## Appendix 5

### Summary of Actions for a Look-Back



## Appendix 6 – Key Contacts

<b>Australian National CJD Registry (ANCJDR)</b> Department of Pathology The University of Melbourne Parkville, Victoria 3052 Telephone: (03) 8344 5868 or (03) 8344 1949 Fax: (03) 8344 4004      Email: <a href="mailto:ANCJD-REG@unimelb.edu.au">ANCJD-REG@unimelb.edu.au</a>	
<b>Australian Government</b> <b>Department of Health and Ageing</b> Office of Health Protection Telephone: +61 2 6289 8951 Fax: +61 2 6289 7100 Email: <a href="mailto:ICG@health.gov.au">ICG@health.gov.au</a> BSE hotline: 1800 200 701	<b>For media inquiries, please contact:</b> Kay McNiece Director Media Unit Department of Health and Ageing Telephone: (02) 6289 5027 Fax: (02) 6289 4044 Mobile: 0412 132 585
<b>Key State and Territory Health Department Contacts</b> All cases of suspect cCJD should be reported immediately to the local Health Department:	
<b>ACT Health</b> Health Protection Service, Communicable Disease Control GPO Box 825 Canberra City ACT 2601 (02) 6205 2155 Email: <a href="mailto:HealthACT@act.gov.au">HealthACT@act.gov.au</a>	<b>WA Health Department</b> PO Box 8172 Perth Business Centre Perth WA 6849 Telephone: (08) 9222 4222
<b>SA Health Department</b> <b>Communicable Disease Control Branch</b> PO Box 6 Rundle Mall Adelaide SA 5000 Telephone: (08) 8226 7177	<b>VIC Department of Human Services</b> 50 Lonsdale Street Melbourne VIC 3000 Telephone: 1300 651 160 Email: <a href="mailto:infectious.diseases@dhs.vic.gov.au">infectious.diseases@dhs.vic.gov.au</a>
<b>NT Department of Health and Community Services</b> PO Box 40596 Casuarina NT 0811 Telephone: (08) 8999 2400	<b>TAS Department of Health and Human Services</b> GPO Box 125 Hobart TAS 7001 Telephone: (03) 6233 3185
<b>NSW Health Department</b> Locked Mail Bag 961 North Sydney NSW 2059 Telephone: (02) 9391 9000 Email: <a href="mailto:NSWhealth@doh.health.nsw.gov.au">NSWhealth@doh.health.nsw.gov.au</a>	<b>QLD Health Department</b> GPO Box 48 Brisbane QLD 4000 Telephone: (07) 3234 0111

Public Health Units in NSW					
Metropolitan Areas			Rural Areas		
Northern Sydney / Central Coast	Hornsby	02 9477 9400	Greater Southern	Goulburn	02 4824 1837
	Gosford	02 4349 4845		Albury	02 6021 4799
South Eastern Sydney / Illawarra	Randwick	02 9382 8333	Greater Western	Broken Hill	08 8080 1499
	Wollongong	02 4221 6700		Dubbo	02 6841 5569
Sydney South West	Camperdown	02 9515 9420	Hunter / New England	Bathurst	02 6339 5601
	Liverpool	02 9828 5944		Newcastle	02 4924 6477
Sydney West	Penrith	02 4734 2022	North Coast	Tamworth	02 6767 8630
	Parramatta	02 9840 3603		Port Macquarie	02 6588 2750
Justice Health Service	Matraville	02 9289 2993		Lismore	02 6620 7500
NSW Department of Health	Nth Sydney	02 9391 9000			
NSW Health website	<a href="http://www.health.nsw.gov.au">www.health.nsw.gov.au</a>				

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