

Creutzfeldt–Jakob disease

Key points

- This document provides recommendations for infection prevention and control procedures to minimise the risk of transmission of Creutzfeldt - Jakob disease (CJD) in health care settings. CJD will be used in this document to refer to all forms of classical Creutzfeldt - Jakob disease.
- Variant CJD (vCJD) is excluded from the scope of this document as vCJD has not been reported in Australia to date. Infection prevention and control issues regarding patients with suspected or confirmed vCJD will be made available on the Department of Health and Ageing website (www.health.gov.au) once vCJD is reported in Australia.
- There is presently no test available to detect CJD infection before the onset of symptoms.
- There is no evidence that CJD can be transmitted through normal social or sexual contact.
- The decision to implement additional procedures for equipment reprocessing is based on a risk assessment (Section 2.4) which incorporates the currently known infectivity of the tissue to which the instrument has been exposed (Section 2.2 and Table 1) and the currently known patient factors (Section 2.3 and Appendix 1 and 2). The additional procedures that may apply as a result of the risk assessment are outlined in Section 3 (and Table 2).
- Although transmission of CJD 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 CJD (the prion) is resistant to routine reprocessing, making the additional procedures outlined in this document essential for the treatment of patients with an identified risk of CJD infection.
- All health care providers and facilities should be familiar with these guidelines and adhere to them, so that patients who may have been exposed to CJD have access to appropriate evidence-based health care without discrimination and disadvantage.

1 Introduction

Creutzfeldt - Jakob disease (CJD) is an invariably fatal human prion disease belonging to the Transmissible Spongiform Encephalopathies (TSEs). These conditions are caused by a pathological accumulation in the brain of an aberrant form (PrP^{Sc}) of a normal cell surface glycoprotein, prion protein (PrP). CJD occurs in familial, sporadic, and acquired (iatrogenic and variant CJD) forms. The familial forms of CJD are autosomal dominant traits associated with mutations in the prion protein gene (PRNP).

The Communicable Diseases Network Australia (CDNA) published guidelines for infection control for classical CJD in 2007, as a revised supplementary chapter (31) of the *Infection Control Guidelines for the Prevention of Transmission of Infectious Diseases in the Health Care Setting*, 2004 edition. These guidelines have now been replaced by the NHMRC *Australian Guidelines for the Prevention and Control of Infection in Healthcare* (2010), which do not include extensive advice on CJD, but instead refer to the CDNA guidelines as the definitive Australian advice on this topic. The CDNA CJD guidelines have now been revised and will be maintained on the Communicable Diseases Network Australia website.

These guidelines provide recommendations for infection prevention and control procedures to minimise the risk of transmission of CJD in health care settings.

The infective agent of CJD (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 procedures outlined in this document essential for treatment of patients with an identified risk of CJD infection. The decision to implement additional procedures for the reprocessing of certain instruments and equipment is based on the currently known infectivity of the tissue to which the instrument or equipment has been exposed (see Table 1) and patient risk factors (refer to Sections 2.3 and 2.4 and Appendices 1 and 2). Provided that the care of the patient is not compromised, alternative diagnostic and management strategies, if suitable and available, can be considered in patients at risk of CJD.

Continual advances in instrument design and reprocessing technology mean that recommendations to minimise the risk of CJD transmission in health care settings should be regularly updated. Health care establishments should ensure that they have the most current version of these guidelines by checking the CDNA website.

Variant CJD (vCJD) is excluded from the scope of this document as vCJD has not yet been reported in Australia. Separate Infection Prevention and 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). If you suspect a patient has vCJD, contact your local State or Territory Health Department immediately.

1.1 Disease Categories

For simplicity, CJD is used to describe all forms of human Transmissible Spongiform Encephalopathies (TSE) except vCJD, including (Collins *et al* 2001, 2004, Brown *et al* 2000, Zerr *et al* 2009):

- 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

1.2 Diagnosis

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

Investigations that may assist in diagnosis of CJD 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) (it is essential that there is minimal red cell contamination of the specimen)
- magnetic resonance imaging (MRI)
- direct amplification of misfolded prion protein (PrP^{Sc}) in the CSF using Real Time-Quaking Induced Conversion (RT-QuIC) (Atarashi *et al* 2011)

2 Assessing the risk

The application of transmission-based precautions to minimise the risk of transmission of CJD is based on a risk assessment. The tissues or body fluids likely to be exposed during a procedure should be classified according to Section 2.2 (and Table 1) and the patient risk category should be identified according to Section 2.3. A risk assessment should then be performed according to Section 2.4. The additional procedures that may apply as a result of the risk assessment are outlined in Section 3 (and Table 2).

2.1 Modes of transmission

Most cases of CJD are sporadic. However, there is evidence of past 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 CJD in the health care setting is exceedingly rare, HCW should be aware of the potential for transmission from patient to patient by contaminated instruments or equipment or via contaminated tissues. There is no epidemiological evidence to indicate that HCW are at an increased occupational risk for CJD. There is no epidemiological evidence that CJD 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).

2.2 Infectivity of human tissues

Table 1 is a guide to the known or predicted infectivity of body tissues and fluids of symptomatic and asymptomatic patients with CJD. This information is based largely on studies of experimentally transmitted CJD 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 1 group the tissues and fluids according to the reprocessing that will be required after contact with these tissues (Brown 1994).

Table 1 Known or predicted infectivity of human body tissues and fluids for CJD

Infectivity category	Tissues	Secretions & excretions
High-infectivity or medium-infectivity ⁽¹⁾ (Higher-infectivity)	Brain Dura mater Pituitary gland Spinal cord Posterior segment of the eye# Cranial and dorsal root ganglia Olfactory epithelium	
Low-infectivity or no detectable infectivity ⁽²⁾ (Low-infectivity)	Cornea ⁽³⁾ Anterior segment of eye@ ⁽³⁾ Kidney Liver Lung Lymph nodes/spleen/ tonsils 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 Ovaries Testes Thyroid gland	CSF Amniotic fluid Faeces Breast milk Nasal mucus Saliva Semen Serous exudate Sweat Tears Urine

#Posterior segment of the eye includes: posterior hyaloid face; retina; retinal pigment epithelium; choroid; subretinal fluid; optic nerve

@ Anterior segment of the eye includes: ocular adnexal tissue including eyelids, periorbital tissue and lacrimal system; conjunctiva; cornea and limbus; iris; crystalline lens; anterior vitreous (excluding the posterior hyaloid face); extra-ocular muscles; ciliary body; sclera (but not if allogeneic sclera used); tissues of the orbit except optic nerve

- (1) 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).
- (2) Low Risk of Transmission (instruments having contact with these tissues and fluids only, do not require additional reprocessing precautions- refer to Appendix 4). Assignment of different organs and tissues to categories of high and low infectivity is chiefly based upon the frequency with which infectivity has been detectable, rather than upon quantitative assays of the level of infectivity, for which data are incomplete. Experimental data include primates inoculated with tissues from human cases of CJD, but have been supplemented in some categories by data obtained from naturally occurring animal TSEs. Actual infectivity titres in the various human tissues other than the brain are extremely limited, but data from experimentally-infected animals generally corroborate the grouping shown in the table.
- (3) Consider the use of single use instruments in known high risk patients, as there has been one definite case of CJD following corneal graft, and one probable case following keratoplasty (Appendix 1, for risk assessment see Section 2.4).

Sources: Modified from: WHO *Guidelines on Tissue Infectivity Distribution in Transmissible Spongiform Encephalopathies* (2006); UK *Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection* (2003); Annex A1: Distribution of TSE infectivity in human tissues and body fluids: updated January 2012

http://www.dh.gov.uk/prod_consum_dh/groups/dh_digitalassets/@dh/@ab/documents/digitalasset/dh_132095.pdf ; UK *Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection* (2003); Annex L: Managing CJD/vCJD risk in ophthalmology: updated January 2011 http://www.loc-net.org.uk/uploaded_files/12073208214303/opt_031_vcjd_disinfection_update.pdf

2.3 Patient risk categories

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

- **High-risk** – people who represent a *definite* risk of CJD transmission (**Appendix 1**). These patients typically report neurological symptoms and display neurological signs of disease;
- **Low-risk** – people who represent a *potential* risk of CJD transmission (**Appendix 2**). These patients may report neurological symptoms or be showing neurological signs or may have an identified risk factor.

In Australia the most common identified risk factor is having received a human pituitary hormone product prior to 1986. Many of these people will have received a “Medical in Confidence” letter from the Chief Medical Officer to provide to their health care practitioner(s) concerning this risk;

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 CJD are likely to have a very low, but unquantifiable risk for CJD. Until further information on the likely risk of these individuals is available, they are conservatively placed in a low risk category. Patients involved as part of a look-back will have received a “Medical in Confidence” letter from the relevant state health department or hospital.

- **Background risk** – the general population who represent no identified increased risk of CJD transmission. The rate of CJD deaths in the general Australian population is 1.15 per million people per year.

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 1). Patients are divided into those with a high risk, those that are considered low risk and those with background risk.

Table 2 Risk assessment matrix

Patient risk category	Procedures involving exposure to higher-infectivity tissues (see Table 1)	Procedures involving exposure to low or no detectable infectivity tissues
High-risk patient	Use additional procedures	Use routine reprocessing procedures ##
Low-risk Patient	Use additional procedures	Use routine reprocessing procedures
Background risk patient	Use routine reprocessing procedures	Use routine reprocessing procedures

See note (3) in Table 1 concerning recommendations relating to surgery involving the cornea or anterior segment of the eye.

Additional procedures (Section 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.

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

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

Questionnaires should be administered to patients by the health care practitioner conducting the procedure that involves higher-infectivity tissues, 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-CJD risk category, the planned procedure may be modified or a process initiated for the implementation of additional procedures for instrument or equipment reprocessing/disposal. Health care establishments should establish systems to ensure that risk assessment, as recommended by this guideline, is undertaken and documented e.g. 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 CJD, patient admission and treatment is not delayed. It is essential that patient care is not compromised and the patient is not discriminated against. To do either is unethical. Any reasons for variations or delays in treatment should be 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- CJD Risk Assessment' is included in Appendix 4.

3 Additional Procedures

Additional procedures 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 procedures that apply to the handling and reprocessing of surgical instruments and diagnostic equipment where CJD may be present are shown in Table 2.

For routine hospital, long-term residential or community care not involving exposure to higher-infectivity tissues, only routine reprocessing procedures are required for the management of CJD patients.

3.1 Reasons for additional reprocessing procedures

The 'prion', which is the infectious agent of CJD, 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. Multiple reprocessing steps will reduce infectivity, but a number of reprocessings which renders instruments completely safe has never been established.

Single use instruments should be used where possible and when use will not compromise patient care. Refer to Section 3.6

3.2 Additional reprocessing procedures

NB These reprocessing procedures are in addition to routine 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 (Fichet *et al* 2004, Jackson *et al* 2005). 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.

Harsh acids and alkalis are not recommended for use in reprocessing due to occupational health issues and potential for damage to instruments and equipment. (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). New enzymatic cleaning products are available, though their effectiveness in deactivating prions requires further assessment.

Instruments and equipment that need to be quarantined or kept for exclusive use in a particular patient and have been exposed to higher-infectivity tissues, or which are to be used exclusively for autopsies on cadavers with confirmed or suspected CJD, 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 or equipment that have been in contact with higher- infectivity tissues in high- or low-risk patients should be separated from other instruments where possible in the operating room to avoid cross-contamination;
- The CJD 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 following surgery until they are reprocessed (for subsequent quarantine or exclusive use in that patient);
- Instruments should be cleaned in anionic detergent prior to further reprocessing. Alternative methods of reprocessing (proteases or alkaline detergents) continue to be actively researched. At the time of writing an enzymatic cleaning solution has been placed on the Therapeutic Goods Administration's Australian Register of Therapeutic Goods (ARTG). However as there is currently insufficient evidence of efficacy of these agents and procedures in the removal of prions, their use is not recommended as an alternative to destruction of instruments used on higher-infectivity tissues in high- or low- risk patients. These products may be incorporated into routine reprocessing for all surgical instruments and equipment.
- In the future, cleaning products and methods may become available which are effective in removing or deactivating prions. However, such products should only be used if placed on the ARTG together with a specific statement confirming that the product or procedure is indicated for the removal or inactivation of prions. Health care establishments should consider this when purchasing new instrument cleaning systems.
- 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;
- Equipment reprocessing staff should wear gloves, fluid-repellent gowns, masks and eye protection at all times when handling higher- infectivity tissues and instruments or equipment 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.

Table 3 Additional Procedures required ONLY for patients identified as high-or-low CJD risk and undergoing procedures involving higher-infectivity tissue

Activity	Additional Procedures
Operating Room Preparation and Set-up	<p>Where possible, schedule patients to allow for appropriate preparation and cleaning following the procedure.</p> <p>Where possible, remove unnecessary equipment and supplies from the operating suite.</p> <p>Where appropriate and it will not present a fire hazard, cover equipment not exposed to higher-infectivity tissue with plastic to protect from splash. Incinerate after use.</p>
Personal Protective Equipment	Wear fluid repellent single use PPE including gloves, gowns and full face shields. Incinerate after use.
Anaesthetic Equipment	Routine management and reprocessing.
Surgical Drapes	All drapes should be single use and incinerated after use.
Tracking of Instruments	HCFs performing procedures exposing higher-infectivity tissue and companies providing loan equipment should have systems in place to track individual reusable items to the level of the individual patient to minimise the number of patients implicated in a look-back.
Instrument Use	<p>Use single-use instruments wherever possible and incinerate OR</p> <p>Reusable instruments should be kept for exclusive use on individual patient. Reprocess separately and quarantine instruments pending determination of risk status. If determined high-or-low: incinerate or keep for the exclusive use of the patient and incinerate on completion of therapy OR place back in circulation if risk found to be background only.</p>
Intra-operative Handling of Instruments	<p>Where possible, separate instruments used on higher-infectivity tissue from other instruments to reduce risk of contamination.</p> <p>Where possible, to prevent tissue residues drying on instruments during surgery, regularly wipe instruments with a moistened radio-opaque pack or keep in a tray/kidney dish covered with a moistened radio-opaque pack.</p>
Reprocessing Instruments (For quarantine or exclusive patient use)	<p>To prevent drying prior to reprocessing immerse instruments contaminated with higher-infectivity tissue in sterile water in a dedicated container after surgery.</p> <p>Reprocess separately.</p> <p>Do not mix with any other instruments or equipment at any stage.</p> <p>Instruments should not be exposed to chemical disinfectants prior to initial cleaning.</p> <p>Steam sterilise at 134°C for 3 minutes.</p> <p>Any item identified as difficult to clean should be destroyed or advice sought from the National CJD Incident Panel.</p>
Quarantine Process	<p>Ensure instruments are separated, reprocessed, contained, labelled and stored in a secure environment pending incineration or return to circulation once risk status determined.</p> <p>Any quarantine system should minimise the risk of accidental re-introduction of potentially infected equipment.</p>
Collection of Specimens	Standard specimen collection, handling and transportation. The specimens should be clearly labelled including a CJD risk alert to laboratory HCWs.
Environmental Cleaning	<p>Routine containment and cleaning procedures apply unless major contamination with higher-infectivity tissue has occurred.</p> <p>Spill-Kits containing either 1M sodium hydroxide (NaOH) or 20,000ppm (free chlorine) sodium hypochlorite should be available in areas of increased risk such as neurosurgery operating rooms, mortuaries and laboratories. Occupational health and safety recommendations and material safety data sheets (MSDS) must be available. Expose area with freshly prepared solution for 1 hour and then rinse with water. Surfaces that cannot tolerate NaOH or sodium hypochlorite should be cleaned using anionic detergent.</p>
Waste Disposal	All items including spent specimens / operative tissue / fluids involved in the case to be disposed of in clinical waste for incineration. Sharps should be disposed of in single-use sharps containers that meet AS 4939.
Endoscopes	<p>Any endoscope⁺ used in a procedure in a high-or-low risk patient where higher-infectivity tissue has been exposed (e.g. ventriculoscope) should be destroyed by incineration or kept exclusively for that patient.</p> <p>In all other situations, endoscopes may be reprocessed using routine processes</p> <p>⁺ Normal nasal endoscope procedures do not reach the olfactory epithelium</p>

3.3 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, equipment and trays will minimise the number of patients implicated in a look-back (Section 4.3), where a patient thought to be at background risk is subsequently diagnosed with CJD.

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

3.4 Quarantine of reusable instruments and equipment used on higher-infectivity tissues

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 used on higher-infectivity tissues should be used if the patient's CJD risk status is not known, including during an investigation by the State or Territory Health Department. Equipment used on higher-infectivity tissues should be quarantined until the risk status is clarified.

If risk clarification determines the patient as background risk for CJD, equipment should be returned to circulation after reprocessing.

If risk clarification determines the patient as low- or high-risk for CJD, equipment used on higher infectivity tissues should be kept for exclusive use on that patient or destroyed by incineration or alternate approved method of medical waste destruction when no longer required.

In some instances, the National CJD Incident Panel (see Section 4.2) may recommend additional reprocessing before instruments or equipment are returned to circulation.

3.5 Destruction of equipment by incineration

Contaminated articles should be placed immediately into the correct clinical waste container for disposal by incineration or alternate approved method of medical waste destruction. Needles, blades and other sharp articles should be placed in non-reusable sharps containers (in accordance with AS 4031:1992 *Non-reusable containers for the collection of sharp medical items used in health care areas*) and destroyed by incineration.

3.6 Single-use surgical instruments

There are a number of single use instruments now available for use in a range of surgical specialties. Careful specification and quality control of instruments are necessary to ensure patient safety.

When using single use instruments for higher risk tissue procedures in lower and higher risk patient groups consider the following:

- Patient care and clinical need should not be compromised. Single use instruments should perform as well as reusable instruments
- Instrument availability
- Cost effectiveness, including appropriate costing required for purchase and disposal
- Possible delays in treatment due to single use instrument availability
- Possible delays in treatment for other patients if reusable instruments must be destroyed or quarantined

It is recommended that surgical staff consider whether instrument sets can be reconfigured and/or single use instruments used, with appropriate segregation and tracking, so that the entire set need not be quarantined or destroyed.

3.7 Environmental cleaning of the operating room

Unless a spill of higher- infectivity tissues has occurred, routine containment and cleaning procedures should be used for the whole operating room, 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 CJD 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 operating room 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.

3.8 Occupational exposure to higher- infectivity tissues

There are no additional requirements following occupational exposure to tissues of a patient with CJD or a patient in the high- or low-risk category.

4 Surveillance

CJD is a notifiable disease in all States and Territories in Australia and is reported to the National Notifiable Diseases Surveillance System (NNDSS). Each State and Territory will have requirements for reporting notifiable diseases, including CJD, and methods for providing advice regarding infection prevention and control issues. See Appendix 6 for State and Territory Health Department contact details.

4.1 Surveillance by the Australian National CJD Registry (ANCJDR)

The Australian Government Department of Health and Ageing (DOHA) established the ANCJDR in 1993, based in the Department of Pathology at the University of Melbourne. The registry assists the DOHA with the ongoing surveillance of CJD cases in Australia, identifies CJD risk factors for population health and provides advice in suspect CJD cases to public health authorities. The contact details of the registry are provided in Appendix 6.

4.2 Surveillance for Adverse Event Management

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

In the event of patients being exposed to instruments that have previously been exposed to higher-infectivity tissues of a patient subsequently found to have CJD, or to the cornea or anterior segments of the eye of a high risk patient, the following should be immediately notified:

- the executive of the health care facility; 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 CJD. The relevant State or Territory Health Department assumes responsibility and is accountable for determining action to be taken, the investigation, instrument or 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 instruments or equipment having direct contact with higher-infectivity tissue (Table 1) have been used in the past on a patient subsequently found to have CJD, the instruments or 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 or equipment will either be destroyed or returned to use following reprocessing. Other instruments or equipment that have not been in contact with a higher-infectivity tissue should not be withdrawn and should continue to be reprocessed using routine methods.

4.3 Look-back

The National CJD Advisory Committee (sometimes referred to as CJD Incident Panel) is available to provide expert advice to inform decisions on the need for a look-back and infection prevention and control measures.

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 facility is responsible for tracing individuals suspected of exposure to instruments or equipment potentially contaminated with CJD.

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 patient privacy and the maintenance of confidentiality of patient details, the way in which information is provided (personal phone communication, face-face, written information), standardised or individualised information and involvement of the media.

People potentially exposed to CJD through the use of human pituitary hormones should have received a letter from the Australian Government informing them of this risk. Patients who have been identified in look-back procedures will also usually be provided a "Medical in Confidence" letter outlining the process and their potential exposure. These people may provide such a letter on admission to hospital.

4.4 Organs and tissues for transplantation

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

- 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 CJD has been specifically excluded
- people who die of dementia
- 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 CJD 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) has an example organ donation referral form.

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

5 Infection Prevention and Control in other settings

5.1 Dentistry and oro-facio-maxillary surgery

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 standard infection prevention and control procedures. In all patients, including higher- and lower-risk patients (Appendix 1 and 2), instruments or equipment in contact with lower infectivity tissues (Table 1) through routine dental procedures can be routinely reprocessed.

Dental work on high- or low-risk patients involving exposure to higher-infectivity tissues should be performed at a facility with HCW who are familiar with CJD specific infection prevention and control procedures.

Instruments or equipment used in oro-facio-maxillary surgical procedures that come into contact with **higher-infectivity** tissues in patients of high- or low- risk should be reprocessed using additional procedures (Table 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 (cranial 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)

5.2 Post Mortem Examinations

Mortuary facilities with staff appropriately trained in CJD infection prevention and 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 adequately funding autopsies for suspected CJD patients and appropriate guidelines and procedures for adequately funding transport of deceased suspected CJD patients to and from autopsy facilities.

Transmission based precautions should be used for autopsies involving exposure to higher infectivity sites in patients with suspected CJD or of high- or low risk, as per Table 2 (Bell and Ironside 1993, Budka *et al* 1995). Disposable instruments or a set of instruments dedicated to suspected CJD patients should be used and **must be kept separate to any instruments used to harvest organs and tissues for donation**. These instruments should be reprocessed using additional reprocessing procedures as per Section 3, Table 2 and Section 3.3.

Removal of the brain with either an electric bone saw or a hand saw should be performed with sufficient containment to effectively capture any aerosol produced.

All tissue samples from higher-infectivity sites should be treated as potentially infectious for CJD until proved otherwise. Tissue or fluid samples should be collected into sealed containers with the CJD 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 autopsy, the body should be sealed in plastic to contain fluid leakage.

Cadavers from high or low-risk patients should not be used for teaching purposes.

5.3 Funeral Industry procedures

Standard disinfection procedures and routine embalming solutions are ineffective against "prions": however, studies show that chemical solutions involving bleach and sodium hydroxide or and physical processes such as autoclaving can inactivate prions.

Autopsies of CJD patients are routinely restricted to the removal of the brain. If the bodies of CJD patients have not undergone a brain autopsy, the transportation, preparation, disinfection, and final disposition can be safely performed when standard precautions are strictly observed. If a brain autopsy has been undertaken, higher level precautions are needed as outlined below.

Transporting:

Using standard precautions and routine personal protective wear, funeral service workers can safely transport the body of a CJD patient for all post-death transfers. It is recommended that the body be placed in a leak proof pouch prior to transport. The bag should be lined with absorbent material to absorb leakage of body fluid. In instances where there is excess fluid, a double bag can be utilized. After transport, all surfaces (i.e. stretchers, cots) should be disinfected as per routine infection prevention and control measures. If leakage occurs during transport, decontamination guidelines should be followed.

Embalming:

Embalming bodies of CJD patients is discouraged.

If embalming and/or reconstruction procedures are needed for transfer overseas, crypt burial or for funereal rites, the following guidelines are strongly recommended (Centers for Disease Control, 2011):

- **For CJD patients who have not been autopsied, embalming can be safely performed.**
 - The body should be placed on a waterproof sheet to collect bodily fluids and disposable instruments should be used whenever possible.
 - All perfusion and washing fluids should be collected in a suitable container for decontamination and disposal.
 - Incision sites should be closed with cyanoacrylates (super glue), wiped with sodium hypochlorite (20,000 ppm available chlorine) and the entire body washed prior to dressing.
 - Routine cosmetic restorative work may also be undertaken using standard infection prevention and control measures.
- **For CJD patients who have been autopsied, embalming and/or reconstruction can be safely performed, if necessary.**
 - Additional infection prevention and control precautions should be undertaken when either viewing or embalming are needed.
 - Special care should be taken to limit fluid leakage when performing any restorative work on a CJD patient.
 - Disposable instruments, masks, safety glasses, gowns, and puncture resistant gloves should be used whenever possible.
 - A plastic sheet with absorbent wadding and raised edges should be placed underneath the head to ensure containment of all fluids, in case of leakage from the cranial cavity.
 - During the reconstruction, the cranial cavity should be packed with absorbent material and tightly sutured to avoid the possibility of fluid leaks from the cranial incision.
 - The entire body should be wiped with sodium hypochlorite (20,000 ppm available chlorine) before dressing.
 - All perfusion and washing fluids should be collected in a suitable container for disposal (incineration) as clinical waste.

Although the use of disposable instruments is preferred, reusable instruments and tools can be reprocessed (cleaned and sterilized) following CJD sterilisation protocols for surgical equipment used on high risk tissues (see Section 3.2 "Additional reprocessing procedures"). Environmental surfaces should be cleaned as outlined in Section 3.7.

Body fluids and chemicals, washing fluids and disposable equipment including cleaning/absorbent cloths should be disposed of according to clinical waste guidelines; i.e. incinerated by a locally licensed incinerator/contractor. Routine non-incineration disposal methods are not adequate.

Casketing & Viewing:

Avoid unnecessary manipulation of the body that would force purging of body fluids and risk opening incision sites. If required, the casket can be lined with an impermeable sheet. An open casket for viewing should not be prohibited. However, **if an autopsy has been performed**, family members of CJD patients can view the deceased after an autopsy and subsequent reconstruction, but should be advised to avoid superficial contact with the body (such as touching or kissing the body).

Cremation & Burial:

There are no special interment or cremation requirements for patients with CJD. Interment of bodies in closed caskets does not present a significant risk of environmental contamination and cremated remains are considered sterile, as the infectious agent does not survive cremation temperatures.

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Appendix 1: Individuals in the high-risk category for CJD

Classification of CJD (Surveillance Definitions)	Clinical signs and risk factors
<p>1 SPORADIC TSE</p> <p>1.1 Definite Neuropathologically/ immunocytochemically confirmed</p> <p>1.2 Probable (refer to clinical signs in adjacent column)</p> <p>1.2.1 Clinical sign I plus at least 2/4 of signs in groups II and III</p> <p>1.2.2 Possible I plus positive 14-3-3 CSF assay</p> <p>1.3 Possible I plus 2/4 of II and duration <2 years</p>	<p>Clinical signs:</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>2 ACCIDENTALLY TRANSMITTED (IATROGENIC) TSE</p> <p>2.1 Definite Definite TSE with a recognised health care acquired risk factor</p> <p>2.2 Probable</p> <p>2.2.1 Progressive predominant cerebellar syndrome in human pituitary hormone recipients</p> <p>2.2.2 Probable TSE with recognised health care associated risk factor</p>	<p>Recognised health care acquired risk factors:</p> <p>Treatment with human cadaver-derived pituitary growth hormone, human cadaver-derived pituitary gonadotrophin or human dura mater graft.</p> <p>Corneal graft in which the corneal donor has been classified as definitely or probably having a human prion disease.</p> <p>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.</p> <p>The relevance of any exposure to disease causation must take into account the timing of exposure in relation to disease onset.</p> <p>This list is provisional, as previously unrecognised mechanisms of human prion disease may occur.</p>
<p>3 GENETIC PRION DISEASES/TSE (includes PRNP mutations associated with CJD, GSS and FFI phenotypes)</p> <p>3.1 Definite</p> <p>3.1.1 Definite TSE <i>and</i> definite or probable TSE in first-degree relative</p> <p>3.1.2 Definite TSE with a pathogenic PRNP mutation</p> <p>3.2 Probable</p> <p>3.2.1 Progressive neuropsychiatric disorder <i>and</i> definite or probable TSE in first-degree relative</p> <p>3.2.2 Progressive neuropsychiatric disorder <i>and</i> pathogenic PRNP mutation</p>	<p>Prion protein gene (PRNP) mutations</p> <p>PRNP mutations associated with GSS neuropathological phenotype:</p> <ul style="list-style-type: none"> • P102L, P105L, A117V, G131V, F198S, D202N, Q212P, Q217R, M232T, 192 bpi <p>PRNP mutations associated with CJD neuropathological phenotype:</p> <ul style="list-style-type: none"> • 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 <p>PRNP mutations associated with FFI neuropathological phenotype:</p> <ul style="list-style-type: none"> • D178N-129M <p>PRNP mutation associated with vascular PRP amyloid:</p> <ul style="list-style-type: none"> • Y145S <p>PRNP mutations associated with proven but unclassified prion disease:</p> <ul style="list-style-type: none"> • H187R, 216 bpi <p>As outlined above, a large number of mutations in the prion protein gene have been described in persons dying of prion disease (Kovacs <i>et al</i> 2002, 2005). In addition to these mutations that appear sufficiently evaluated to be classed as causally linked to the development of prion disease, a small number are less well characterized and therefore have a tentative association with human</p>

	<p>prion disease; for example:</p> <p>Mutations associated with neuropsychiatric disorder but not proven prion disease:</p> <ul style="list-style-type: none">• I138M, G142S, Q160S, T188K, M232R, 24 bpi, 48 bpi, 48 bpi + nucleotide substitution in other octapeptides <p>PRNP mutations without clinical and neuropathological data:</p> <ul style="list-style-type: none">• T188R, P238S
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Note: The following people are also classified as being at high risk: carriers of disease-linked mutations of the PRNP, and persons in whom the PrP gene has not been sequenced but who have two or more first or second degree relatives with CJD (including GSS or FFI). People who have had the PRNP 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 CJD

<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 if <1 year duration and where professional review is unable to assign a high-risk status or a background risk status.</p>
<p>All genetically related members of any family in which there is a strong family history (two or more first or second -degree relatives¹) of dementia or neurological illness, and in which affected individuals have not been competently and completely assessed, specifically for CJD.</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 CJD are likely to have a very low, but unquantifiable risk for CJD 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>

¹ First degree relative: parent, sibling, or child

Second degree relative: grandparent, grandchild, uncle, aunt, nephew, niece, half-sibling

Appendix 3: Classical Creutzfeldt - Jakob Disease (CJD) Risk Assessment Tool

INTRODUCTION

It is recommended this questionnaire be completed well ahead of planned elective surgery so precautions can be put into place if needed. 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 CJD?

Yes	No
-----	----

Q2. Has the patient had two or more first or second-degree relatives with CJD? (It is important to know about any relatives with CJD, but having a single affected relative with sporadic CJD does not place the patient in a low- or high-risk category.)

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 CJD or shown you a 'medical in confidence letter' regarding their risk for CJD?

Yes	No
-----	----

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

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

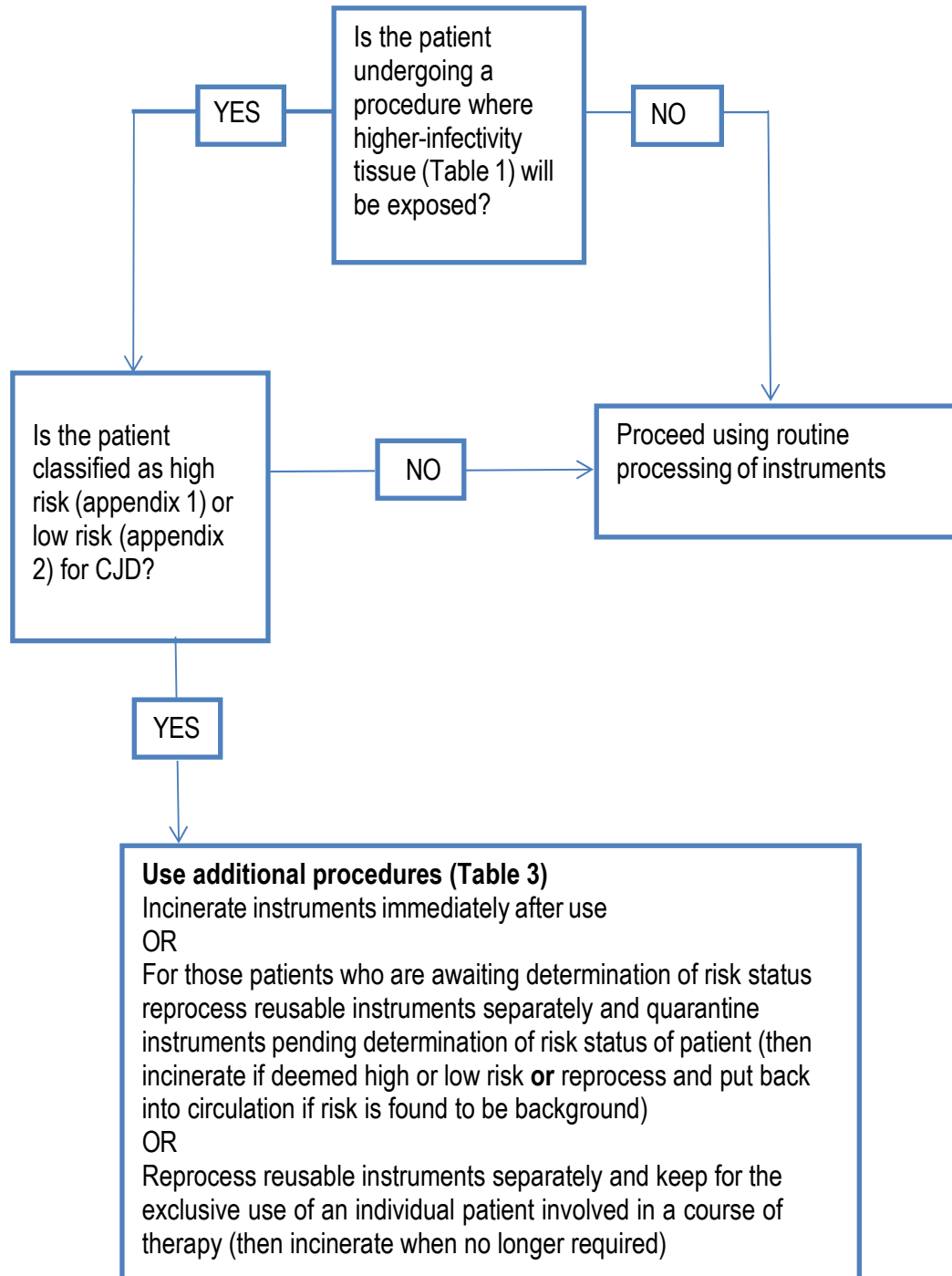
Name of the Health Care Practitioner

Signature

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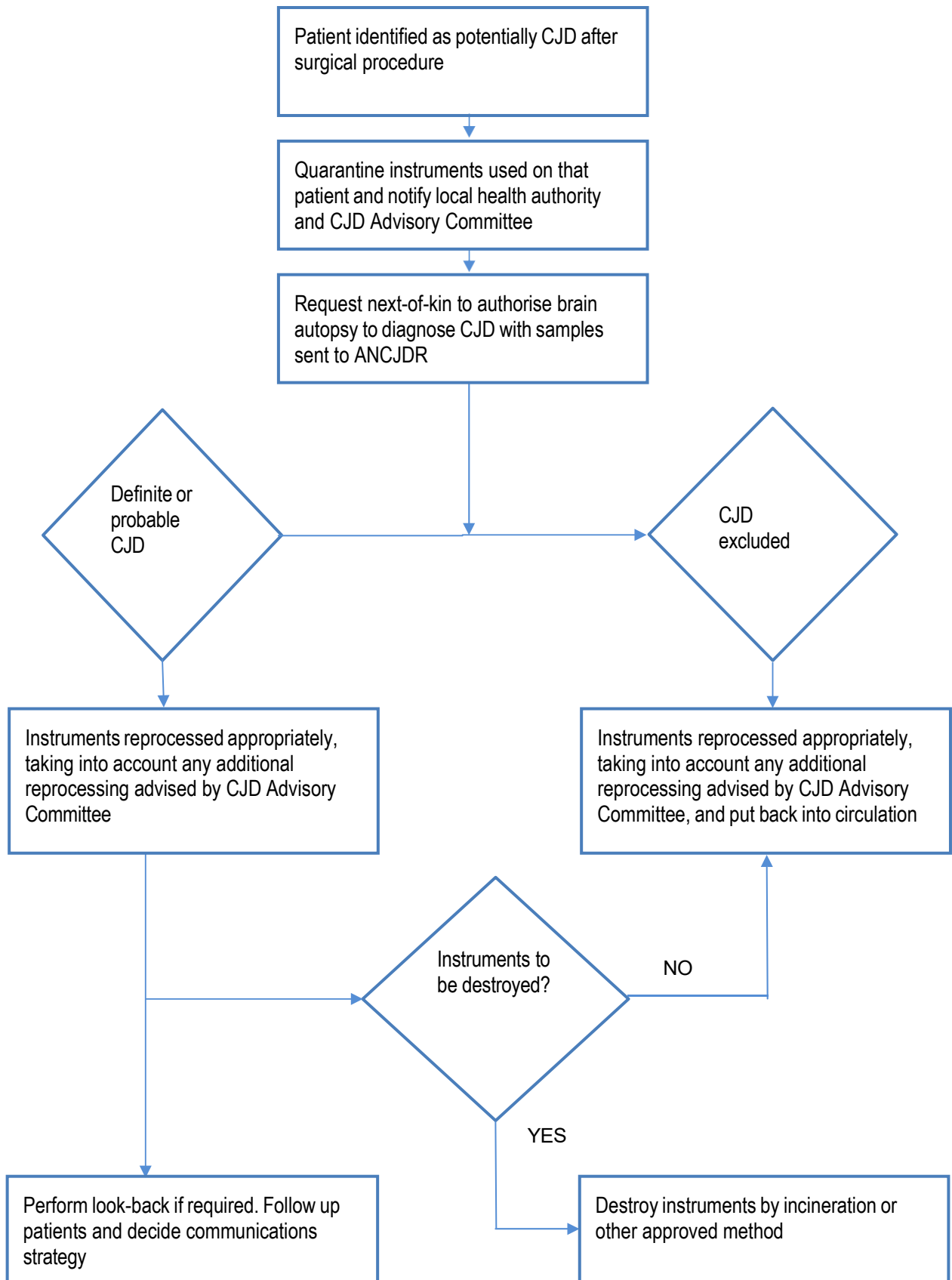
Appendix 4: Summary of Actions for a Surgical Procedure – CJD Risk Assessment

This flow chart describes what the procedure for treating surgical instruments if a patient is classified as having an increased risk of CJD contamination.



Appendix 5: Summary of Actions for a Look-Back

This flow chart summarises the procedure to follow if a patient has undergone a surgical procedure that has involved surgical instruments that have potentially been exposed to CJD



Appendix 6 – Key Contacts

Australian National CJD Registry (ANCJDR)

Department of Pathology
The University of Melbourne
Parkville, Victoria 3052
Telephone: (03) 8344 5868 or (03) 8344 1949
Fax: (03) 8344 4004 Email: ANCJD-REG@unimelb.edu.au

Australian Government Department of Health and Ageing

Office of Health Protection
Telephone: +61 2 6289 2726
Fax: +61 2 6289 2600

For media inquiries, please contact:

Director
Media Unit
Department of Health and Ageing
Telephone: (02) 6289 5027
Fax: (02) 6289 4044
Mobile: 0412 132 585

Key State and Territory Health Department Contacts

All cases of suspect CJD should be reported immediately to the local Health Department:

ACT Health. Health Protection Service, Communicable Disease Control

GPO Box 825
Canberra City ACT 2601
(02) 6205 2155
Email: HealthACT@act.gov.au

WA Health Department

Communicable Disease Control Directorate
PO Box 8172
Perth Business Centre
Perth WA 6849
Telephone: (08) 9388 4868

SA Health Communicable Disease Control Branch

PO Box 6
Rundle Mall
Adelaide SA 5000
Telephone: 1300 232 272
Email: cdc@health.sa.gov.au

VIC Department of Health

50 Lonsdale Street
Melbourne VIC 3000
Telephone: 1300 651 160
Email: infectious.diseases@health.vic.gov.au

NT Department of Health and Community Services

PO Box 40596
Casuarina NT 0811
Telephone: (08) 8999 2400

TAS Department of Health and Human Services

GPO Box 125
Hobart TAS 7001
Telephone: 1800 671 738

NSW Health Department

Locked Mail Bag 961
North Sydney NSW 2059
Telephone: (02) 9391 9000
Email: NSWhealth@doh.health.nsw.gov.au

QLD Health Department

GPO Box 48
Brisbane QLD 4000
Telephone: (07) 3234 0111

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