



OPERATIONAL DIRECTIVE

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Subject: **Classical Creutzfeldt-Jakob Disease (cCJD) Risk Assessment, Screening and Management**

Compliance with this Operational Directive is mandatory.

INTRODUCTION

This Operational Directive describes 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 document as vCJD has not been reported in Australia to date. This Directive should be read in conjunction with the guidelines contained in 'Chapter 31 Classical Creutzfeldt-Jakob disease' revised December 2007 and released by the Commonwealth Department of Health and Ageing.

1. BACKGROUND

Creutzfeldt-Jakob disease is a rare disease of the central nervous system which results in death, after a relatively rapid course of muscle weakness and dementia for which there is no known cure. The term Classical Creutzfeldt-Jakob disease (cCJD) is used to describe all forms of human transmissible spongiform encephalopathies (TSE) affecting humans except vCJD, including:

1. Sporadic CJD	2. Inherited CJD	3. Acquired CJD
	2.1 Familial CJD. 2.2 Gerstmann-Sträussler-Scheinker disease (GSS). 2.3 Fatal Familial Insomnia (FFI).	3.1 Healthcare associated CJD. 3.2 Kuru.

Although transmission of cCJD in the healthcare setting is very rare, Health Care Workers (HCW) need to be aware of the potential for transmission by instruments or equipment contaminated by higher-infectivity tissues. The infective agent of cCJD (the prion) is resistant to routine reprocessing, making the additional precautions outlined in this document essential for the treatment of patients with an identified risk of cCJD. The decision to implement additional precautions for equipment reprocessing is based on a risk assessment which incorporates the current known infectivity of the tissue to which the instrument/equipment has been exposed and patient risk factors.

For routine hospital, long term residential, palliative or community care not involving exposure to higher-infectivity tissues, routine reprocessing procedures and standard precautions are all that is required for the management of suspected cCJD patients.

2. DIAGNOSIS

There is currently no minimally invasive test available to detect cCJD infection before the onset of symptoms. A pre-symptomatic period exists 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, biopsy is not recommended as a routine procedure to confirm clinical suspicion of cCJD. Methods which may assist in the diagnosis of cCJD and excluding other causes of subacute dementia in symptomatic patients include:

- Electroencephalograph (EEG);
- Presence of protein 14-3-3 in cerebrospinal fluid; and
- Imaging techniques, e.g. computerised tomography and magnetic resonance imaging.

3. MODES OF TRANSMISSION

The majority of cases of cCJD are sporadic. However, although transmission of cCJD in the healthcare setting is very rare, there is evidence of healthcare associated transmission through the use of neurosurgical instruments contaminated with central nervous system (CNS) tissue and through contaminated tissue implants or products (dura mater grafts, corneal grafts and pituitary products).

There is no epidemiological evidence to indicate HCWs are at an increased occupational risk for cCJD, or that cCJD can be transmitted through normal social or sexual contact, mother-to-child transmission or via blood or blood products.

4. INFECTIVITY OF HUMAN TISSUE

The known or predicted infectivity of body tissues and fluids of symptomatic and asymptomatic patients with cCJD are listed in **Appendix 1**.

5. PATIENT RISK CATEGORIES

It is international convention to define three risk categories that reflect the theoretical and demonstrable risks of transmitting cCJD.

High-risk	Low-risk	Background risk
People who represent a definite risk of cCJD transmission. These patients are generally showing neurological symptoms.	People who represent a potential risk of cCJD transmission. These patients may be showing neurological symptoms or may have an identified risk factor.	The general population who represent no identified increased risk of cCJD transmission.

The characteristics used to identify individuals who may pose a risk of transmitting cCJD are detailed in **Appendix 2**.

6. RISK ASSESSMENT AND MANAGEMENT

6.1 Assessment

All patients undergoing surgical or diagnostic procedures involving higher-infectivity tissue (neurosurgery, spinal cord surgery, ophthalmic surgery, pituitary surgery) shall have their cCJD risk category determined prior to the procedure. A questionnaire template to assist with risk assessment for cCJD is attached as **Appendix 3**.

The patient risk assessment questionnaire shall be administered to patients by the medical practitioner performing the procedure, prior to consent being obtained. The completed questionnaire is to be included in the patient medical record.

6.2 Management

Health care facilities (HCF) must ensure processes are in place to ensure the risk assessment, where recommended (**Appendix 4**), is undertaken and actioned appropriately. In the event a patient is identified to be in a high-or-low-cCJD risk category undergoing a procedure involving higher-infectivity tissue, each HCF is to have an action plan in place to ensure patient admission and treatment is not unnecessarily delayed. There is a need to ensure patient care is not compromised and that any reasons for variations or delays in treatment are clearly communicated to the patient.

The planned procedure may require modification or initiation of processes for the implementation of additional precautions for equipment reprocessing. All HCWs involved in the care of the patient, equipment reprocessing or environmental cleaning are to be fully informed of the proposed additional precautions being implemented.

Additional precautions are **only** to be implemented as outlined in Table 1 when:

- the diagnostic or therapeutic procedure performed involves the exposure of **higher-infectivity tissue**. (Appendix 1) **AND**
- the patient is identified as **high-or-low risk** (Appendix 2).

Table 1: Risk Assessment Matrix

Patient Risk Categories	Procedures Involving Exposure to Higher-Infectivity Tissue	Procedures Involving Exposure to Lower-Infectivity Tissue
HIGH RISK	Additional Precautions Required	Standard Precaution & Routine Reprocessing Equipment
LOW RISK	Additional Precautions Required	Standard Precaution & Routine Reprocessing Equipment
BACKGROUND RISK	Standard Precaution & Routine Reprocessing Equipment	Standard Precaution & Routine Reprocessing Equipment

7. ADDITIONAL PRECAUTIONS

The additional precautions required for identified high-and-low cCJD risk patients having diagnostic or therapeutic procedures involving higher-infectivity tissue are described in **Appendix 5**.

8. INSTRUMENT TRACKING

HCFs and companies that provide loan, demonstration or trial equipment for use in procedures exposing higher-infectivity tissues shall 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, where a background risk patient is subsequently diagnosed with cCJD.

9. NOTIFICATION AND ADVERSE EVENT MANAGEMENT

Creutzfeldt-Jakob disease has been scheduled as a notifiable disease in all Australian States and Territories and on receipt of confirmatory pathology it must be notified to the Communicable Disease Control Directorate (CDCD). Notification is also indicated where a strong clinical suspicion for CJD exists.

Since there is no test to reliably detect cCJD prior to 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 this event, the equipment having direct contact with higher-infectivity tissue should be identified via instrument tracking systems and quarantined. Immediate notification to the Executive of the HCF and to the Director of CDCD is to occur.

The National CJD Incident Panel, established by the Australian Government, is available to provide expert advice in the event of an adverse event involving cCJD. The Director of CDCD is to be notified of all possible adverse events and will assume responsibility for the investigation, equipment management, patient risk assessment and the scope of a look-back investigation if required.

10. INFECTION CONTROL IN OTHER SETTINGS

10.1 Dentistry

Oro-facio-maxillary surgical procedures that come into contact with higher-infectivity tissues in patients of high-or-low risk shall be treated with additional precautions. These procedures would include:

- Major oral surgical procedures such as maxillectomy with orbit enucleation (optic nerve).
- Injection of the trigeminal ganglion (potential brain tissue, central nerve exposure).
- Oral surgical cancer procedures combining a neurosurgical approach (potential brain tissue, central nerve exposure).

In all patients, instruments in contact with lower-infectivity tissue (Appendix 1) through routine dental procedures can be routinely processed. An appropriate medical history should be taken for all patients. Dental work on high-or-low risk patients involving exposure to higher infectivity tissues should be performed at an establishment with HCWs who are familiar with cCJD additional precautions.

11. POST MORTEM EXAMINATIONS

Additional precautions are to be used for post mortems involving exposure to higher infectivity tissue in patients with suspected cCJD or those assessed as high-or-low risk. 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.

REFERENCES

1. Communicable Disease Network Australia (2004). Infection Control Guidelines for the Prevention of Transmission of Infectious Diseases in the Health Care Setting. Chapter 31: Classical Creutzfeldt -Jacob Disease. Revised December 2007.
<http://www.health.gov.au/internet/main/publishing.nsf/Content/icg-guidelines-index.htm>
2. Australian Government, Department of Health and Ageing. The Use of Human Pituitary Hormones in Australia and Creutzfeldt - Jakob disease. Updated 16 November 2007.
<http://www.health.gov.au/internet/main/publishing.nsf/Content/health-pubhlth-strateg-phi-index.htm>
3. Australian Government, Department of Health and Ageing. Creutzfeldt - Jakob disease Fact sheet.
http://www.health.gov.au/internet/main/publishing.nsf/Content/icg-guidelines-cjd_factsheet.htm
4. National Institute of Neurological Diseases and Stroke. Creutzfeldt - Jakob disease Information Page. <http://www.ninds.nih.gov/disorders/cjd/cjd.htm>
5. Queensland Health. Guideline: Management of Classical Creutzfeldt - Jakob disease. August 2007. Centre for Healthcare Related Infection Surveillance & Prevention. Queensland Government.
6. NSW Health. Creutzfeldt - Jakob disease (CJD) Transmission During High Risk Surgical Procedures. Safety Notice SN: 003/07. 19 March 2007. Quality and Safety Branch NSW Department of Health.

ADDITIONAL RESOURCES

The Australian National CJD Registry (ANCJDR) – Melbourne

Phone: (03) 8344 1949

Email: ancjd-reg@unimelb.edu.au

Visit: <http://ancjdr.path.unimelb.edu.au/>

The ANCJDR is under contract to the Commonwealth to assist with the ongoing surveillance of TSE in Australia. The registry will act as a resource to assist with clarification of cases into high-low-background risk for cCJD. They offer diagnostic services to enhance ante-mortem diagnostics; the 14-3-3 protein CSF test and genetic testing. The ANCJDR is a World Health Organisation reference centre for human TSE.

CJD Support Group Network

Phone: 1800 052 466 (Toll Free)

Email: contactus@cjdsupport.org.au

Visit: <http://www.cjdsupport.org.au>

The CJD Support Group Network is funded by the Department of Health and Ageing. It is contracted to assist and support families affected by CJD and people who are 'at risk of CJD' in Australia.

The National Pituitary Hormones Advisory Council - Canberra

Phone: 1800 802 306 (0900–1700 EST)

They provide a central contact point for hormone recipients who wish to obtain further information and to keep hormone recipients informed of relevant issues via the HPH newsletter and the departmental web-site with both to be maintained until 2010.

Dr Peter Flett

ACTING DIRECTOR GENERAL

Known or Predicted Infectivity of Body Tissues and Fluids for cCJD

Infectivity Category	Tissues	Secretions
HIGH or MEDIUM Infectivity ⁽¹⁾ (Higher Infectivity)	<ul style="list-style-type: none"> • Brain • Dura mater • Pituitary gland • Spinal cord • Retina • Optic nerve • Cranial & dorsal root ganglia • Olfactory epithelium (<i>not normally encountered in routine nasal or sinus surgery</i>) 	
LOW or NO DETECTABLE Infectivity ⁽²⁾ (Lower-infectivity)	<ul style="list-style-type: none"> • Cornea⁽³⁾ • Anterior chamber of eye⁽³⁾ • Kidney • Liver • Lung • Lymph nodes / spleen • Placenta • Uterus • Adipose tissue • Adrenal gland • Blood & blood products • Bone marrow • Gingival tissue • Heart muscle • Intestine • Peripheral nerve • Prostate • Skeletal muscle • Testes • Thyroid gland • Trigeminal ganglia 	<ul style="list-style-type: none"> • CSF • Amniotic fluid • Faeces • Breast milk • Nasal mucous • 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)

- (1) Considerable risk of Transmission - instruments having contact with these tissues **may** require additional reprocessing precautions
- (2) Low Risk of Transmission – instruments having contact with these tissues and fluids only **do not** require additional precautions
- (3) It is recommended that single use instruments be used in known high risk patients

Classification of High-risk and Low-risk patients for cCJD *

High-risk patients
<p>1. Sporadic TSE</p> <ul style="list-style-type: none"> • Patients with neuropathologically or immunocytochemically confirmed CJD. • Patients with clinically suspected CJD i.e. rapidly progressive dementia, typical EEG and at least two clinical signs attributable to CJD. • Patients with possible cCJD and a positive 14-3-3 CSF assay. • Patients with rapidly progressive dementia, at least two clinical signs attributable to CJD and duration < 2 years. <p>2. Accidentally transmitted TSE</p> <ul style="list-style-type: none"> • Definite TSE with a recognised health care associated risk (i.e. exposure to surgical instruments contacted with higher-infectivity tissue in a definite or probable CJD case; corneal graft recipient in which the donor has been classified as definite or probable TSE; recipients of cadaver-derived human pituitary hormones or dura mater homografts). • Progressive predominant cerebellar syndrome in human pituitary hormone recipients. • Probable TSE with a recognised health care associated risk factor. <p>3. Genetic TSE</p> <ul style="list-style-type: none"> • Definite TSE and definite or probable TSE in 1st degree relative. • Definite TSE with a pathogenic prion protein (PRNP) mutation. • Progressive neuropsychiatric disorder and definite or probable TSE in first degree relative. • Progressive neuropsychiatric disorder and pathogenic PRNP mutation. • Carriers of disease linked mutations of the PrP gene. • Patients in whom the PrP gene has not been sequenced but have two or more first degree relatives with cCJD (including GSS or FFI).
Low-risk patients
<ul style="list-style-type: none"> • People with an undiagnosed progressive neurological illness of < 1 year's duration, with or without dementia awaiting the outcome of a professional review to assign a high-risk or background risk status or those patients for whom a determination cannot be following professional review. • Patients undergoing a diagnostic brain biopsy for progressive brain disease. • Patients undergoing neurosurgical investigations for a progressive disorder that includes dementia. • 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. • Recipients of cadaver-derived human pituitary hormones (growth hormone and gonadotrophins) before 1986. • 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. • Individuals who have been contacted by a health authority as part of a look-back procedure from exposure to surgical instruments previously used on higher infectivity tissues in patients later found to have contracted cCJD.

* Table Adapted from Communicable Disease Network Australia (2004). Infection Control Guidelines for the Prevention of Transmission of Infectious Diseases in the Health Care Setting: Section 31 (Revised December 2007). This document is to be referenced for complete classification when assigning risk category. <http://www.health.gov.au/internet/main/publishing.nsf/Content/icg-guidelines-index.htm>

Patient Questionnaire - Risk Assessment Tool

Please use Patient Identification Label or Block Print

**IDENTIFICATION OF POTENTIAL CREUTZFELD
JAKOB DISEASE (CJD) RISK**

SURNAME	MRN
GIVEN NAME	
DOB	SEX
DOCTORS NAME	

Admitting Medical Practitioner to Complete

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:

- Brain, pituitary or dura mater
- Cranial and dorsal root ganglia
- Spinal cord
- Eye (Retina / Optic nerve)
- Olfactory epithelium (not normally encountered in routine nasal or sinus surgery)

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.

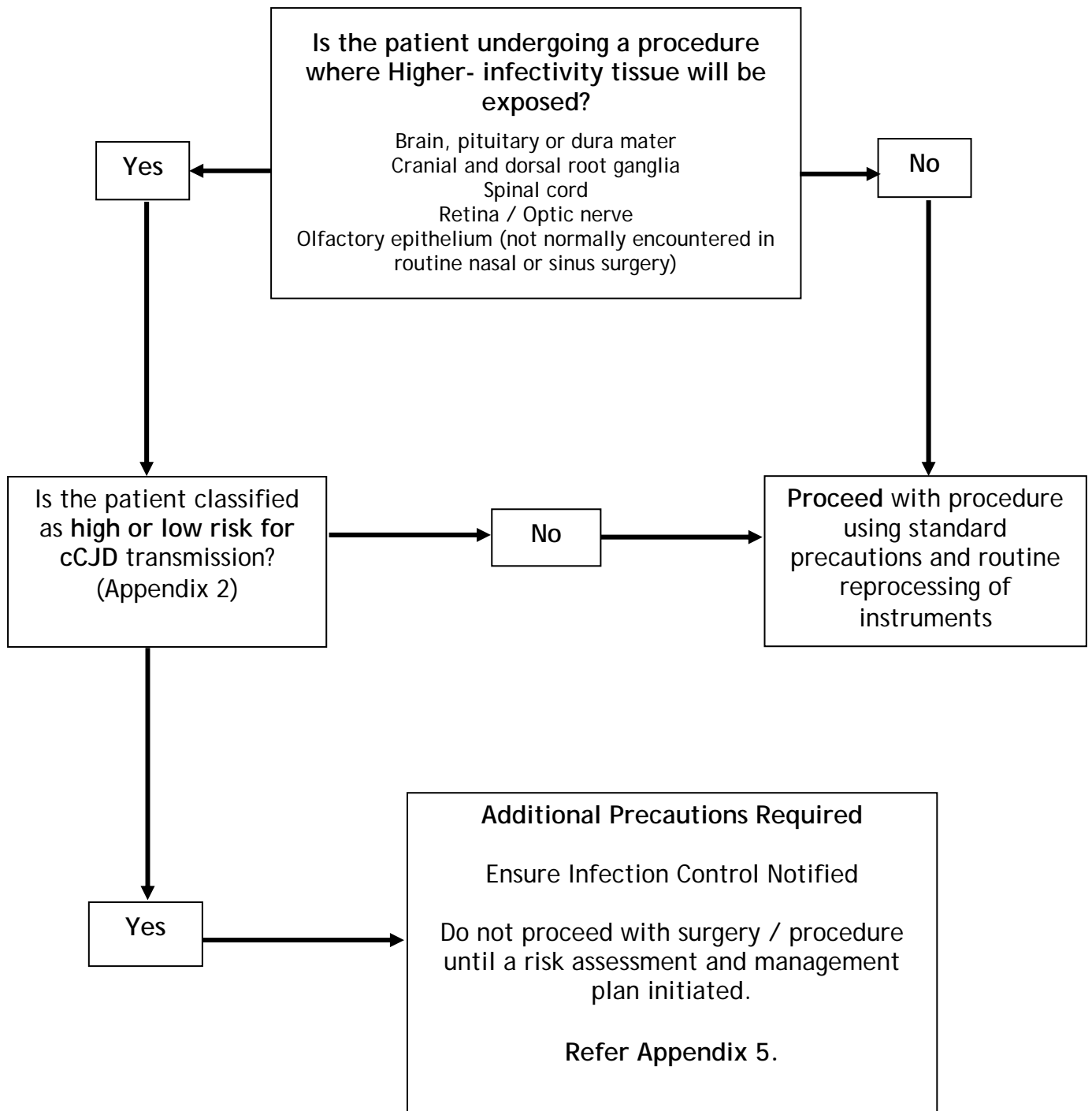
To be completed by patient’s medical officer:

	QUESTIONS	YES	NO
1	Do you think the patient may have cCJD?		
2	Has the patient had a first degree relative with cCJD?		
3	Does the patient have an unexplained progressive neurological illness of less than 12 months?		
4	Does the patient have a history of receiving human pituitary hormone for infertility or human growth hormone for short stature prior to 1986?		
5	Has the patient previously had surgery on the brain or spinal cord that included a dura mater graft prior to 1990?		
6	Has the patient been involved in a ‘look-back’ for cCJD or shown you a ‘medical in confidence’ letter regarding their risk for cCJD?		

If the patient answers ‘**YES**’ to any of the above questions, please contact **<insert name of responsible person>** for further advice. Additional precautions may need to be implemented for re-processing of equipment. **DO NOT** proceed to surgery prior to obtaining this advice.

Advice Sought from: _____	<Print Name>
Admitting Doctor: _____	<Print Name>
Signature: _____	Date: ____/____/____

Risk Assessment Flow Chart to Identify if Additional Precautions Required



Additional precautions required **ONLY** for patients identified as high-or-low cJD risk and undergoing procedures involving higher-infectivity tissue

Process	Additional Precautions
Preparation and Set-up	<ul style="list-style-type: none"> › Schedule patients to allow for appropriate preparation and cleaning. › Remove unnecessary equipment and supplies from the operating suite. › Cover equipment not exposed to higher-infectivity tissue with plastic to protect from splash. Incinerate after use.
Personal Protective Equipment	<ul style="list-style-type: none"> › Wear fluid repellent single use PPE including gloves, gowns and full face shields. Incinerate after use.
Anaesthetic Equipment	<ul style="list-style-type: none"> › Routine management and reprocessing.
Surgical Drapes	<ul style="list-style-type: none"> › All drapes to be single use and incinerate after use.
Tracking of Instruments	<ul style="list-style-type: none"> › HCFs performing procedures exposing higher-infectivity tissue and companies providing loan equipment shall 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	<ul style="list-style-type: none"> › Use single-use instruments wherever possible and incinerate OR › Non-single use instruments and incinerate OR › 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.
Intra-operative Handling of Instruments	<ul style="list-style-type: none"> › Separate instruments used on higher-infectivity tissue from general instruments to reduce risk of contamination. › To prevent drying prior to reprocessing immerse instruments contaminated with higher-infectivity tissue in sterile water in a dedicated container.
Reprocessing Instruments (For quarantine or exclusive patient use)	<ul style="list-style-type: none"> › Reprocess separately. › Do not mix with any other instruments or equipment at any stage. › Instruments are not to be exposed to chemical disinfectants prior to initial cleaning. › Clean with an anionic detergent and water using soft brush. › Ultrasonic cleaners and automated washers should not be used in preparatory cleaning. › Steam sterilise at 134°C for 3 minutes. › Any item identified as difficult to clean shall be destroyed.
Quarantine Process	<ul style="list-style-type: none"> › Ensure instruments are separated, reprocessed, containerised, labelled. and stored in a secure environment pending incineration or return to circulation once risk status determined. › Any quarantine system must minimise the risk of accidental re-introduction of potentially infected equipment.
Collection of Specimens	<ul style="list-style-type: none"> › Standard specimen collection, handling and transportation. The specimens should be clearly labelled including a CJD risk alert to laboratory HCWs.
Environmental Cleaning	<ul style="list-style-type: none"> › Routine containment and cleaning procedures apply unless major contamination with higher-infectivity tissue has occurred. › Spill-Kits containing either 1M sodium hydroxide (NaOH) or 20,000ppm (free chlorine) sodium hypochlorite are to be available in areas of increased risk such as neurosurgery operating rooms, mortuaries and laboratories. OS&H 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.
Waste Disposal	<ul style="list-style-type: none"> › All items including spent specimens / operative tissue / fluids involved in the case to be disposed of in clinical waste for incineration. Standard sharps disposal to occur.
Endoscopes	<ul style="list-style-type: none"> › Any endoscope⁺ used in a procedure in a high-or-low risk patient where higher-infectivity tissue has been exposed (e.g. ventriculoscope) shall be destroyed by incineration or kept exclusively for that patient. › In all other situations, endoscopes may be reprocessed using routine processes ⁺ Normal nasal endoscope procedure do not reach the olfactory epithelium