



## OPERATIONAL CIRCULAR

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**Subject:            CREUTZFELDT-JAKOB DISEASE (CJD) SCREENING, RISK ASSESSMENT  
                         AND MANAGEMENT**

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*This Operational Circular is designed to advise Department of Health staff on Departmental policies and procedures.*

### 1. BACKGROUND

Creutzfeldt-Jakob Disease (CJD) 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.

### 2. SOURCE AND TRANSMISSION OF INFECTION

The incidence of CJD in Australia is 1.5 cases per million population per year. Of these cases 91% occur sporadically with no recognisable pattern of transmission. Health care associated (iatrogenic) CJD has been linked with neurosurgical instruments contaminated with neurological tissue and through tissue implants or products (dura mater grafts, corneal grafts, human pituitary products). Globally approximately 5% – 10% of CJD cases present as familial cases which usually develop because of inherited mutations of the prion protein gene.

Experience to date indicates that the highest risk of health care associated transmission occurs when the Central Nervous System is exposed in a patient with CJD. There is no evidence that the disease can be transmitted through normal social or sexual contact or through blood or blood products or by vertical transmission.

Variant CJD (vCJD); the human form of bovine spongiform encephalopathy, or “mad cow disease” has not been reported in Australia. It has been determined that the risk of transmitting vCJD in Australia in the course of health care delivery is extremely remote.

### 3. SYMPTOMS OF CJD

A symptom free incubation period of more than 30 years may occur for CJD that occurs sporadically. For iatrogenic CJD, the symptom free incubation period can range from 18 months to 40 years with an average of 15 years based on international experience. Latrogenic, familial and sporadically occurring CJD all have similar clinical presentations.

The early symptoms of CJD can be vague and there are no firm diagnostic tests to confirm exposure or diagnose CJD until symptoms are well advanced. Onset of symptoms is typically after the age of 60. Diagnosis is made by clinical and neuropathological examination as the illness progresses with the results suggesting the disease. 90% of patients die within one year of diagnosis. Examination of brain tissue by biopsy or autopsy is the only way to obtain a definitive diagnosis of CJD.

Symptoms may include:

- Confusion or disorientation, which rapidly advances to dementia.
- Personality changes.
- Behavioural changes.
- Weakness or loss of balance and muscle control, causing difficulty walking.
- Muscle spasms.
- Visual symptoms such as double vision or blindness.

However, most people with these symptoms do NOT have CJD.

#### 4. RISK ASSESSMENT AND MANAGEMENT OF PATIENTS FOR NEUROSURGICAL, SPINAL AND OPHTHALMOLOGY PROCEDURES

The recommended approach for managing the risk of health care associated transmission of CJD is to identify individuals who have the potential to pose a risk in the health care setting and manage these individuals under conditions that prevent disease transmission. The *National Infection Control Guidelines (2004)*<sup>1</sup> indicate that it is international convention to define two risk categories that reflect the theoretical and demonstrable risks of transmitting CJD:

**Higher-risk** — people who represent a definite risk of CJD transmission; and

**Lower-risk** — people who represent a potential risk of CJD transmission.

##### 4.1 Screening to identify the risk

Institutions should develop a pre-operative questionnaire/risk assessment tool to identify higher risk patients for CJD who are undergoing **neurosurgical, and ophthalmological (posterior segment)** procedures, which involve tissue with the highest risk of infectivity for CJD. See example of a questionnaire (Appendix 4).

**NB:** *The National Infection Control (2004)*<sup>1</sup> *Guidelines* recommend that additional precautions and additional sterilising or reprocessing are indicated for **higher risk patients** undergoing surgery involving tissue classified as low infectivity or no detectable infectivity, i.e. **general surgery**. The rationale is that lymphoid tissue is involved in a range of procedures performed on these tissues. Also, although there is no known human infectivity of blood, there is still uncertainty as to whether blood components carry the infectious prions in humans.

Also, **endoscopes** and other fiberoptic scopes cannot be adequately cleaned and reprocessed to destroy prions and using these instruments on patients in the CJD **higher risk** category should be avoided and alternative diagnostic strategies considered.

##### 4.2 Managing the risk

All institutions should have a notification plan when higher and lower risk patients for CJD are identified, or suspected awaiting confirmation of suspicion. At the least, the following personnel should be notified to determine the course of action and the need for additional CJD precautions:

- The local infection control professional.
- The Communicable Disease Control Directorate.

(1) Determine if the patient is higher risk or lower risk or no risk. See Table 1 - Appendix 1 and preoperative questionnaire - Appendix 4.

- (2) Determine if the procedure to be performed involves high infectivity tissues, low infectivity tissues or no detectable infectivity of tissues. See Table 2 – Appendix 2.
- (3) Use the flow charts to determine course of action for higher risk and lower risk patients. See Figure 1 – Appendix 3.

**NB: No additional precautions are required for patients identified to be at no risk for CJD regardless of the tissues involved in the procedure.**

#### **4.3 Additional precautions for CJD**

The infective agent of CJD (a prion) is very resistant to inactivation and therefore additional precautions are required in relation to the management of the instruments, the theatre environment and staff and, laboratory procedures where specimens are transported and manipulated. Standard precautions are adequate for all other routine health care interactions.

Each institution should develop protocols addressing:

1. **Additional precautions for CJD and additional sterilisation or reprocessing methods for CJD and disposal of instruments for incineration.** See Figure 1 – Appendix 3.
2. **Quarantining of instruments** - the initial cleaning and sterilisation, storage and labelling of quarantined instruments awaiting the outcome of a suspected higher or lower risk case of CJD. Also, the course of action for **quarantined** instruments when the risk status is clarified.
3. **Tracking** of reusable instruments used on high infectivity tissues.
4. **Endoscopes** and other **fiberoptic scopes** and certain ophthalmic and optometric equipment that **cannot be adequately reprocessed** by methods that inactivate prions.

For information on additional precautions for CJD refer to:

Communicable Disease Network Australia (2004). Infection Control Guidelines for the Prevention of Transmission of Infectious Diseases in the Health Care Setting. <http://www.health.gov.au/internet/wcms/publishing.nsf/Content/icg-guidelines-index.htm>.

#### **5. Australian National CJD Registry – Melbourne** **Phone: (03) 8344 1946**

This is a CJD surveillance unit which is bound by confidentiality rules regarding disclosure of information on specific cases. The registry will act as a resource to assist with clarification of cases into high risk or low risk for CJD. Neurosurgeons can also refer tests (e.g. spinal fluid protein) to the Pathology Department – (03) 8344 5868.

#### **6 Pituitary Hormone Hotline - Canberra** **Phone: 1800 802 306 (0900–1700 EST)**

This hotline can be rung to confirm whether a patient received cadaver-derived pituitary hormones before 1986. Recipients can also be referred to a counselling service by phoning this number.

## References

1. Communicable Disease Network Australia (2004). Infection Control Guidelines for the Prevention of Transmission of Infectious Diseases in the Health Care Setting. <http://www.health.gov.au/internet/wcms/publishing.nsf/Content/icg-guidelines-index.htm>  
Accessed January 2004.
2. Australian Government, Department of Health and Ageing. The Use of Human Pituitary Hormones in Australia and Creutzfeldt-Jakob Disease. <http://www.health.gov.au/internet/wcms/publishing.nsf/Content/health-publth-strateg-phi-index.htm>  
Accessed 17 February 2005.
3. Australian Government, Department of Health and Ageing. Creutzfeldt-Jakob Disease Fact sheet. [http://www.health.gov.au/internet/wcms/publishing.nsf/Content/icg-guidelines-cjd\\_factsheet.htm](http://www.health.gov.au/internet/wcms/publishing.nsf/Content/icg-guidelines-cjd_factsheet.htm)  
Accessed 17 February 2005.
4. National Institute of Neurological Diseases and Stroke. Creutzfeldt-Jakob Disease Information Page. <http://www.ninds.nih.gov/disorders/cjd/cjd.htm>  
Accessed 17 February 2005.

Dr John de Campo  
**ACTING DIRECTOR GENERAL**

**SUPERCEDED**

TABLE 1: Identification of patient risk for CJD

Lower risk patients	Higher risk patients
<ul style="list-style-type: none"> <li>• People with undiagnosed progressive neurological illness of &lt; 1years duration, with or without dementia who are awaiting the outcome of assessments.</li> <li>• Patients undergoing a diagnostic biopsy for progressive brain disease.</li> <li>• Patients undergoing any neurological investigations for a progressive brain disease that includes dementia.</li> <li>• Known recipients of cadaver-derived human pituitary hormones (growth hormone and gonadotropins) before 1986.</li> <li>• Known 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.</li> <li>• Patients with a strong family history (two or more first-degree relatives) of dementia or neurological illness in which individuals have not been competently and completely assessed neurologically, specifically for CJD.</li> </ul>	<ul style="list-style-type: none"> <li>• Patients with confirmed CJD.</li> <li>• Patients with clinically suspected CJD i.e. symptoms of CJD and results of medical investigations are consistent with CJD or patient has iatrogenic risk factors.</li> <li>• People with probable genetically acquired CJD, i.e.               <ul style="list-style-type: none"> <li>— (i) progressive neuropsychiatric disorder in first-degree relatives of people with confirmed or probable genetically acquired (familial) CJD.</li> <li>— (ii) progressive neuropsychiatric disorder in carriers of pathogenic prion protein mutations.</li> </ul> </li> <li>• Carriers of disease-linked pathogenic prion protein mutations.</li> <li>• People in whom the prion protein gene has not been sequenced and who have two or more first-degree relatives with CJD.</li> </ul>

**For further details of the risk categories and the prion protein mutations refer to:**

Communicable Disease Network Australia (2004). Infection Control Guidelines for the Prevention of Transmission of Infectious Diseases in the Health Care Setting.  
<http://www.health.gov.au/internet/wcms/publishing.nsf/Content/icg-guidelines-index.htm>

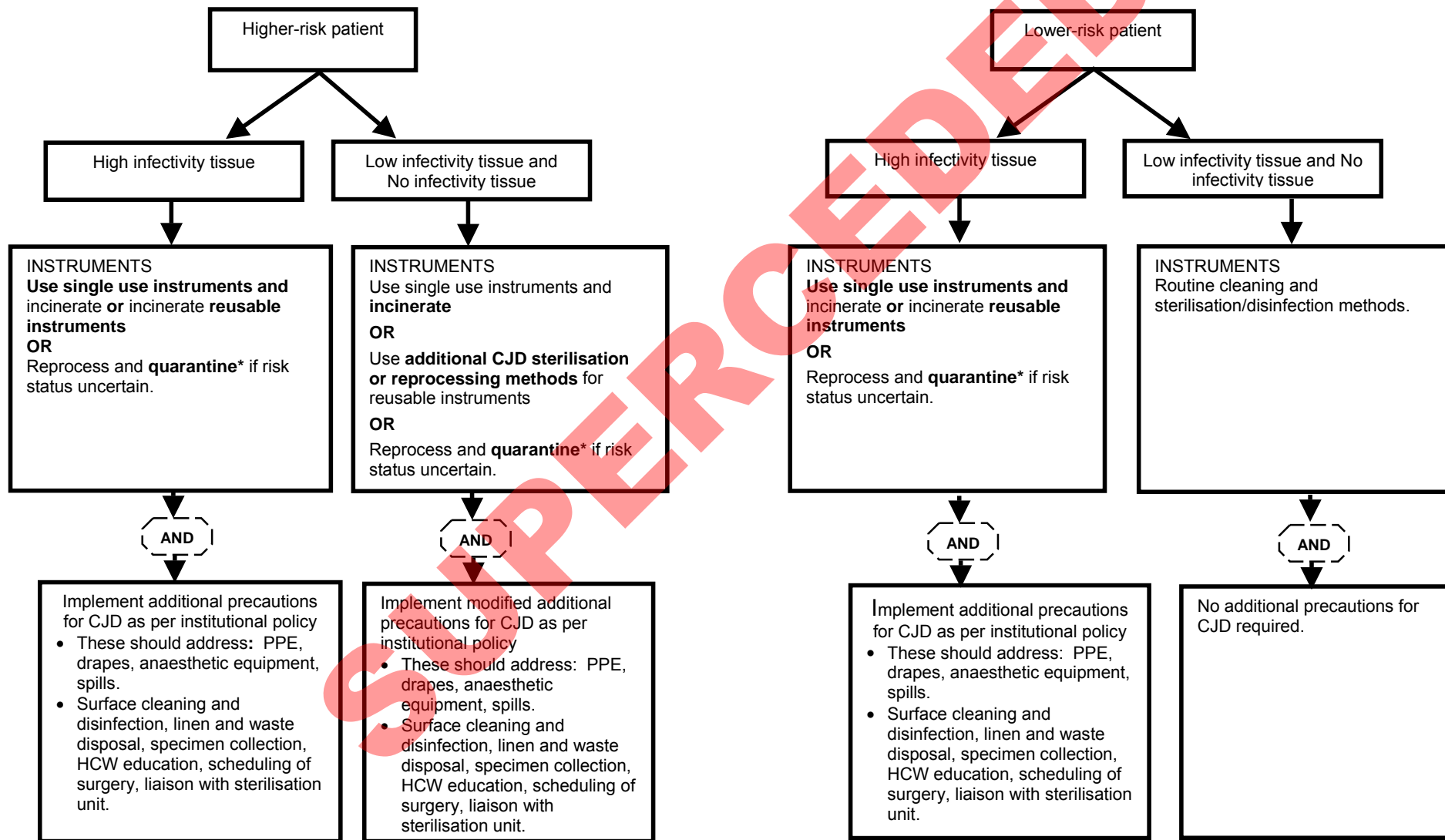
Section 31.9.1 and 31.9.2.

TABLE 2:

DEMONSTRATED OR PREDICTED INFECTIVITY OF HUMAN BODY TISSUES AND FLUIDS FOR CJD		
Infectivity Category	Tissues	Secretions and Excretions
<b>High infectivity</b> (demonstrated or predicted to be consistently infectious)	<ul style="list-style-type: none"> <li>• Brain</li> <li>• Spinal cord</li> <li>• Eye (retina and optic nerve)</li> <li>• Pituitary</li> <li>• Dura mater</li> </ul>	
<b>Low infectivity</b> (demonstrated or predicted to be infectious, but not consistently)	<ul style="list-style-type: none"> <li>• Eye (cornea and anterior chamber)</li> <li>• Kidney</li> <li>• Liver</li> <li>• Lung</li> <li>• Lymph nodes/spleen</li> <li>• Placenta</li> <li>• Uterus</li> <li>• Dorsal root ganglia</li> <li>• Trigeminal ganglia</li> </ul>	CSF
<b>No detectable infectivity</b> (have not been demonstrated to be infectious)	<ul style="list-style-type: none"> <li>• Adipose tissue</li> <li>• Adrenal gland</li> <li>• Blood</li> <li>• Bone marrow</li> <li>• Cartilage and connective tissue</li> <li>• Gingival tissue</li> <li>• Heart muscle</li> <li>• Intestine</li> <li>• Peripheral nerve</li> <li>• Prostate</li> <li>• Skin</li> <li>• Skeletal muscle</li> <li>• Testes</li> <li>• Thyroid gland</li> </ul>	<ul style="list-style-type: none"> <li>• Faeces</li> <li>• Milk</li> <li>• Nasal mucous</li> <li>• Saliva</li> <li>• Semen</li> <li>• Serous exudate</li> <li>• Sweat</li> <li>• Tears</li> <li>• Urine</li> </ul>

**Figure 1: RISK MANAGEMENT OF HIGHER AND LOWER RISK PATIENTS FOR CJD REQUIRING SURGICAL AND INVASIVE PROCEDURES**

**NOTE:** \*Clean, sterilise and store quarantined instruments as per institution's protocol pending determination of risk status. If a CJD risk is identified then reprocess or incinerate according to the patient risk and infectivity of tissue. If no risk is identified then instruments may be returned to circulation.



**APPENDIX 4**  
**Example Only**

Attach patient addressograph label

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

**For pre-operative patients undergoing neurosurgical  
and ophthalmological (posterior segment) procedures**

**To be completed by patient's medical officer:**

	QUESTIONS	YES	NO
1.	Has specialist neurological assessment and investigations raised the possibility of CJD?		
2.	Does the patient have a history of undiagnosed progressive neurological illness of less than 1 year's duration?		
3.	Has the patient received cadaver-derived human pituitary hormones (growth hormone and gonadotropins) before 1986? Telephone the Pituitary Hormone Hotline – 1800 802 306, to access a register of recipients.		
4.	Has the patient received dura mater homografts or had transdural neurosurgery before 1990?		

**If "YES" is answered to one or more of these questions please contact:**

for further advice

Signature of admitting doctor: \_\_\_\_\_ Date: \_\_\_\_ / \_\_\_\_ / \_\_\_\_

Printed name: \_\_\_\_\_