

**Infection Control Guidelines**  
**Classic Creutzfeldt-Jakob Disease in Canada**  
**Quick Reference Guide**

In December 2004, the Public Health Agency of Canada (PHAC) convened a special meeting of the Advisory Committee on Infection Prevention and Control for Creutzfeldt-Jakob Disease, to review and revise as necessary the 2002 Infection Control Guideline for Classic Creutzfeldt-Jakob Disease in Canada.

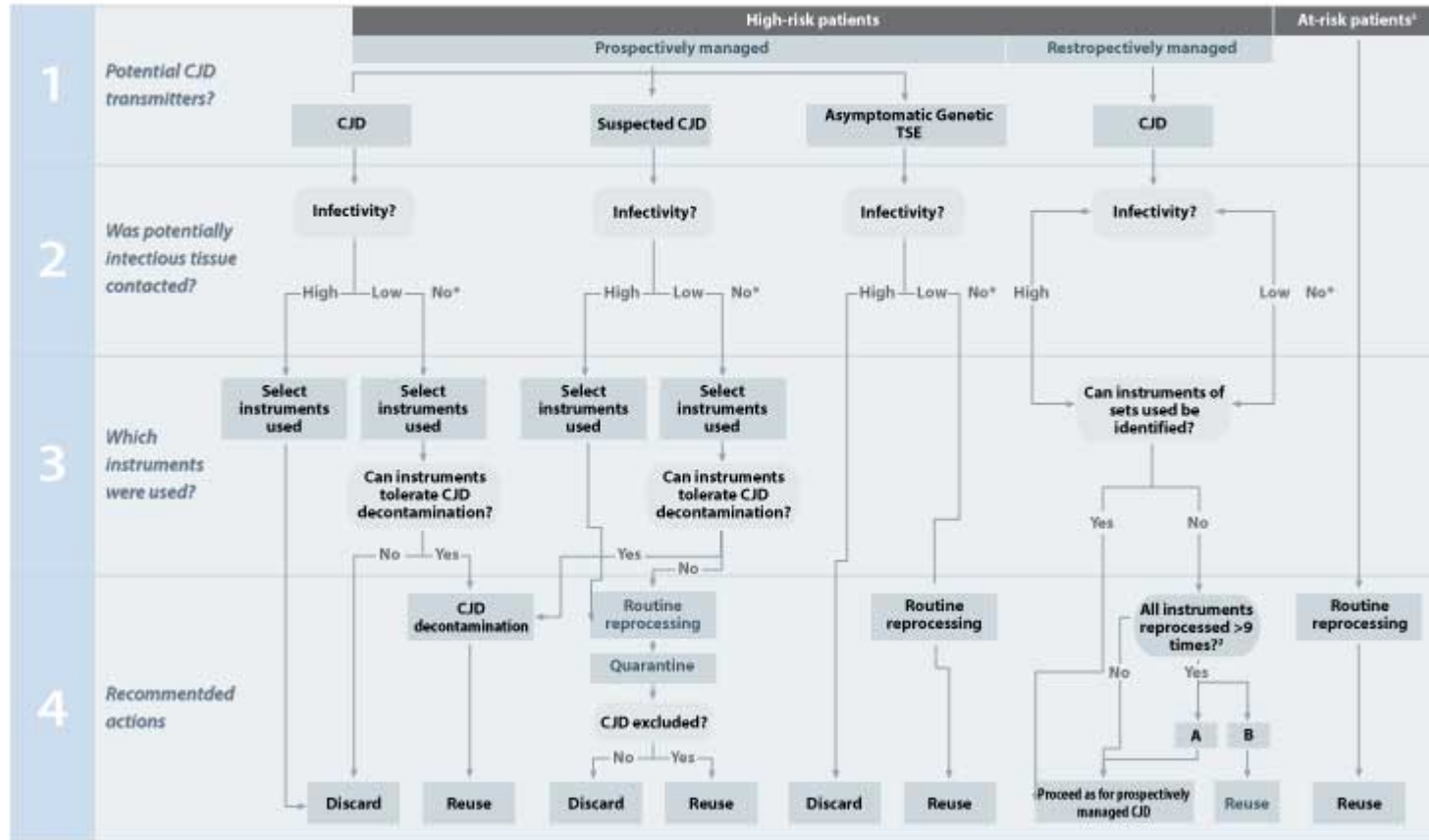
This meeting was preceded by a PHAC International CJD Scientific meeting where the most up-to-date scientific information regarding CJD and its iatrogenic transmission were reviewed.

This document represents clarification and revision of components of the 2002 Guideline based on information available at the time of the 2004 PHAC International CJD Scientific Meeting. See the 2002 Guideline for complete recommendations.

The most effective, safe, and efficient means to prevent iatrogenic transmission of CJD is to identify high risk patients before an invasive procedure, in order to implement the required infection prevention and control measures, and to have a system for instrument tracking (Section 3).

# Quick Reference Guide

Risk Assessment Tool  
 Recommendations for managing instruments used on CJD patients  
 Decision algorithm-graphic version



For explanations of each of the four steps in the above graphic, including definitions, see the corresponding numbered sections in the text below.

<sup>1</sup>No detected infectivity tissue. No CJD precautions. Reprocess as usual.

<sup>2</sup>2 – 9 times: no evidence that it removes all risk.

<sup>3</sup>3 – Dura mater, human pituitary hormone, cornea transplants

# Decision algorithm (textual version)

## High-risk CJD patients managed prospectively

CJD <sup>†</sup>	
<b>Instruments that contacted: †††</b>	<b>Action to be taken: ††††</b>
<b>High-infectivity tissue<sup>††</sup></b>	Discard
<b>Low-infectivity tissue<sup>††</sup></b>	<p><i>Can the instruments tolerate CJD decontamination?</i></p> <p>If yes ---- CJD decontamination &amp; Reuse</p> <p>If no ---- Discard</p>
<b>No-infectivity tissue<sup>††</sup></b>	Routine reprocessing & reuse
Suspected CJD <sup>†</sup>	
<b>Instruments that contacted: †††</b>	<b>Action to be taken: ††††</b>
<b>High-infectivity tissue<sup>††</sup></b>	<p>Routine reprocessing on their own &amp; Quarantine</p> <p><i>Is diagnosis of CJD excluded?</i></p> <p>If yes ---- Reuse</p> <p>If no ---- Discard</p>
<b>Low-infectivity tissue<sup>††</sup></b>	<p><i>Can the instruments tolerate CJD decontamination?</i></p> <p>If yes ---- CJD decontamination &amp; Reuse</p> <p>If no ---- Routine reprocessing on their own &amp; Quarantine</p> <p><i>Is diagnosis of CJD excluded?</i></p> <p>If yes ---- Reuse</p> <p>If no ---- Discard</p>
<b>No-infectivity tissue<sup>††</sup></b>	Routine reprocessing & Reuse
Asymptomatic Genetic TSE <sup>†</sup>	
<b>Instruments that contacted:†††</b>	<b>Action to be taken:††††</b>
<b>High-infectivity tissue<sup>††</sup></b>	Discard
<b>Low/No-infectivity tissue<sup>††</sup></b>	Routine processing and reuse

† See 1. Is the patient a potential CJD transmitter? p.6

†† See 2. Was infectious tissue contacted? p.7

††† See 3. Which instruments were used? p.8

†††† See 4. Action definitions. p.9

## High-risk CJD patients managed retrospectively

CJD <sup>†</sup>	
Instruments that contacted:	Action to be taken: <sup>††††</sup>
High/Low-infectivity tissue <sup>††</sup>	<p><i>Can specific instruments or sets be identified?</i></p> <p>If yes ---- Proceed as for prospectively managed CJD</p> <p>If no ---- <i>Were instruments reprocessed &gt;9 times?</i></p> <p style="padding-left: 40px;">If yes ---- Proceed as for prospectively managed CJD (option A) or Reuse (option B)</p> <p style="padding-left: 40px;">If no ---- Proceed as for prospectively managed CJD (option A)</p> <p>The CJD Incidents Panel (UK) consultation paper concluded that most instruments that have gone through ten cycles of use and decontamination are unlikely to pose a significant risk of cross-transmission. This is based on scenario modeling using the risk assessment for transmission of variant CJD endorsed by the Spongiform Encephalopathy Advisory Committee.</p> <p>There is however no experimental data to confirm these modeling conclusions. Given this information and the significant cost associated with discarding large numbers of surgical instruments the PHAC Infection Control Guidelines Steering Committee felt that this guideline should reflect uncertainty in this area and allow health care facilities option A or B. This decision should consider whether the instruments can be adequately reprocessed.</p>
No-infectivity tissue <sup>††</sup>	Continue to reuse

---

† See 1. Is the patient a potential CJD transmitter? p.6

†† See 2. Was infectious tissue contacted? p.7

†††† See 4. Action definitions. p.9

## At-risk patients

Recipients of human pituitary hormone, duramater grafts and cornea transplants <sup>†</sup>	
<b>Instruments that contacted:</b>	<b>Action to be taken:<sup>†††</sup></b>
<b>High-infectivity tissue<sup>††</sup></b>	Routine reprocessing & Reuse
<b>Low/No-infectivity tissue<sup>††</sup></b>	Routine reprocessing & Reuse

In this update of the 2002 guideline, incineration or CJD decontamination is no longer recommended for the high-infectivity tissues of at-risk patients.

---

<sup>†</sup> See 1. Is the patient a potential CJD transmitter? p.6

<sup>††</sup> See 2. Was infectious tissue contacted? p.7

<sup>†††</sup> See 4. Action definitions. p.9

# 1. Is the patient a potential CJD transmitter?

## High-risk patients

Patients considered to be at high risk of transmitting CJD iatrogenically are those diagnosed, prospectively or retrospectively, with:

- CJD — either confirmed, probable, or possible CJD, familial CJD, GSS, or FFI depending on pathological, laboratory, and clinical evidence and following the Surveillance Definitions for Classic CJD\*
- Suspected CJD — undiagnosed, rapidly progressive dementia and CJD not ruled out
- Asymptomatic genetic TSE — asymptomatic member of a family with familial CJD, Gerstmann-Sträussler-Scheinker (GSS), or fatal familial insomnia (FFI) as determined by genetic testing or having a first-degree relative with a genetic TSE, or by having two or more first-degree relatives with classic CJD

Step 4 outlines appropriate procedures for managing instruments that have been in contact with high-risk patients, depending on the potential infectiousness of the tissue contacted.

To minimize the risk of transmitting CJD, elective procedures in high-risk patients (involving high-risk or low-risk tissues) should be well justified and carefully planned in advance.

## At-risk patients

The following patients are at risk of iatrogenic CJD:

- Recipients of a dura mater graft (until 1992 for Lyodura grafts, until 1997 for Tutoplast grafts)
- Recipients of human pituitary hormone treatment (either growth or gonadotrophin)
- Recipients of a corneal graft originating in a jurisdiction that does not require graft donors to be screened for neurological disease
- Patients who have been exposed, via contact with instruments, to high-risk tissue in a confirmed CJD patient

The working group considers, after examining the available evidence, the risk of transmission via instruments used on at-risk, asymptomatic patients is negligibly low, and therefore recommends such instruments be routinely decontaminated and then reused.

Screening procedures should be performed to identify high-risk patients, and not to identify at-risk patients. A patient who self-identifies as being at-risk should be evaluated clinically for evidence of CJD.

---

\* See 2002 Guideline, Appendix II, p.59 for Surveillance Definitions for Classic CJD, Possible CJD and Probable CJD.

## 2. Was infectious tissue contacted?

The procedures recommended for managing instruments used on high-risk patients depend on the potential infectivity of the tissue contacted. Using evidence from animal studies and reports of iatrogenic exposure, human tissue *is classified* into three categories, according to its risk of transmitting CJD. This information is subject to change as further information becomes available.

High infectivity	No detected infectivity
<ul style="list-style-type: none"> <li>▪ Brain</li> <li>▪ Dura mater</li> <li>▪ Pituitary gland</li> <li>▪ Posterior eye (including optic nerve and retina)</li> <li>▪ Spinal cord</li> <li>▪ Cranial and spinal cord ganglia (including dorsal root ganglia and trigeminal ganglia)</li> <li>▪ CSF*</li> </ul>	<ul style="list-style-type: none"> <li>▪ Adipose</li> <li>▪ Adrenal gland</li> <li>▪ Blood</li> <li>▪ Bone marrow</li> <li>▪ Breast Milk</li> <li>▪ Feces</li> <li>▪ Heart muscles</li> <li>▪ Intestine</li> <li>▪ Nasal mucous</li> <li>▪ Peripheral nerve</li> <li>▪ Prostate</li> <li>▪ Saliva</li> <li>▪ Semen</li> <li>▪ Serous exudates</li> <li>▪ Skeletal muscle</li> <li>▪ Skin</li> <li>▪ Sweat</li> <li>▪ Tears</li> <li>▪ Testis</li> <li>▪ Thyroid gland</li> <li>▪ Urine</li> </ul>
Low infectivity	
<ul style="list-style-type: none"> <li>▪ Anterior eye (includes cornea)</li> <li>▪ Dental neurovascular tissue/dental pulp</li> <li>▪ Kidney</li> <li>▪ Liver</li> <li>▪ Lung</li> <li>▪ Lymph nodes</li> <li>▪ Placenta</li> <li>▪ Spleen</li> </ul>	

In this update of the 2002 guideline anterior and posterior eye have been differentiated in terms of infectivity and cranial and spinal cord ganglia have been classified as high infectivity tissues.

\* Note that contact with CSF necessarily implies contact with dura mater, a high-infectivity tissue, and should be so managed.

### 3. Which instruments were used?

It is recommended to:

- Limit as much as possible the number of instruments used for any procedure
- Use disposable rather than reusable instruments whenever possible and especially when contacting high-infectivity tissue
- When using reusable instruments, choose, whenever possible, those that can tolerate the rigors of CJD decontamination
- Track use of reusable instruments

#### Instrument identification

Without detailed information as to which reusable instruments were in contact with potentially infectious tissue, the only way to eliminate all risk of iatrogenic transmission is to discard all potentially contaminated instruments. This is obviously wasteful.

Without such information, the opportunity to reduce the risk of transmission by instruments already in circulation — a risk to which some patients have already been exposed — is lost.

To reduce or eliminate such risk without waste, it is strongly recommended all reusable instruments be tracked. Technology is available now for tracking instruments, though not widely used, and more effective technology is being developed.

Some sample tracking measures are listed below in order, roughly, of increasing effectiveness and decreasing feasibility. Immediate adoption of the most effective feasible measures is recommended, and the addition of even more effective methods as soon as these become feasible. Facilities that have not yet established a full tracking system (measure 1) should, **at a minimum**, segregate those instruments used on high-risk tissue (measure 2).

1. Identify — for example, by means of a colour code or a bar code — those sets of reusable instruments used only on
  - Brain and neuro/ortho spine
  - Retinal/posterior eye

Keep these sets separate from other sets.

2. Within a set of instruments, identify those instruments used only on high-infectivity tissues. Keep these specific instruments separate from the others.

3. Identify — for example, by means of a bar code — each individual instrument and the set to which it belongs. Associate records of the use of this individual instrument with records of the patients on whom it was used.

## 4. Action Definition

We recommend some combination of the following measures to manage or reduce the risk of iatrogenically transmitting CJD infection through reused instruments.

### Discard

To discard means to assure an instrument cannot possibly transmit infection to another patient. Incineration is the most unambiguous means of doing so. Note, however, there is evidence that prions can survive very high temperatures.

Where incineration is not available, an acceptable alternative is CJD decontamination (see below) followed by disposal in landfill.

### CJD decontamination

We recommend, where appropriate, a combined method of CJD decontamination in four steps:

1. Clean thoroughly
2. Soak in 1N sodium hydroxide (NaOH) for 1 hour
3. Thoroughly rinse
4. Sterilize in a prevacuum-method autoclave at 134°C for 60 minutes

Acceptable substitutions for steps 2 and 4 include:

2. Substitute 2% NaOCl (20,000 ppm available chlorine) for sodium hydroxide (NaOH)
4. Sterilize in a prevacuum-method autoclave at 134°C for 18 minutes rather than 60 minutes

### Notes

- Instruments made of high-quality stainless steel can tolerate CJD decontamination using NaOH
- Instruments that contain plastic or electronic devices, such as bronchoscopes, cannot tolerate CJD decontamination
- Instruments that contain both steel and other metals, and particularly aluminum, should never be exposed to NaOH

Ideally, decontamination procedures shown to deactivate prions would be used on all instruments no matter which patients they were used on or what tissues they contacted. Unfortunately, the only available procedures for which there is evidence of effective deactivation cannot be routinely used as they are potentially damaging to many instruments. We do not believe, based on the evidence, that autoclaving at 134°C for 18 minutes by itself suffices to deactivate prions, and hence do not recommend this as a routine procedure to prevent CJD transmission.

### Routine reprocessing

Clean and sterilize in the recommended (CSA-specified) manner.

## **Quarantine**

After routinely reprocessing separately from other instruments (see Decision algorithm. p. 3,4 and 5), store instruments in dry conditions. Do not reuse unless a diagnosis is made that eliminates the possibility that the patient on whom the instruments were used had CJD. A confirmed diagnosis other than CJD, either clinical or pathological, or a postmortem examination excluding CJD, is required to take instruments out of quarantine. A brain biopsy that is negative for CJD, in the absence of a confirmed alternate diagnosis, does not suffice to take instruments out of quarantine.

## 5. References

- (1) Asher D. *Infection control and decontamination of TSE agents: A US view.* Presentation Public Health Agency of Canada International CJD Scientific Meeting. Ottawa. December 2004.
- (2) Boutoleau C, Guillon B, Martinez F, Vercelletto M, Faure A, Feve JR. *Iatrogenic Creutzfeldt-Jakob disease subsequent to dural graft: persisting risk after 1987.* Eur J Neurol 2003; 10:521-523.
- (3) Brooke FJ. *Reviewing the Australian CJD infection control guidelines.* Presentation Public Health Agency of Canada International CJD Scientific Meeting. Ottawa. December 2004.
- (4) Brooke FJ, Boyd A, Klug GM, Masters CL, Collins SJ. *Lyodura use and the risk of iatrogenic Creutzfeldt-Jakob disease in Australia.* Med J Austral 2004; 180:177-181.
- (5) Brown P, Preece M, Brandel J-P, Sato T, McShane L, Zerr I, et al. *Iatrogenic Creutzfeldt-Jakob disease at the millennium.* Neurology 2000; 55:1075-1081.
- (6) Bryant G. *UK CJD incidence panel.* Presentation Public Health Agency of Canada International CJD Scientific Meeting. Ottawa. December 2004.
- (7) Health Canada. *Infection control guidelines for classic Creutzfeldt-Jakob disease (CJD) in Canada. Part of the Infection Control Guidelines Series.* Canada Communicable Disease Report , 1-84. 2002.  
<http://www.phac-aspc.gc.ca/publicat/ccdr-rmtc/02vol28/28S5/index.html>
- (8) de Pedro Cuesta J. *Surgery and sporadic Creutzfeldt-Jakob disease.* Presentation Public Health Agency of Canada International CJD Scientific Meeting. Ottawa. December 2004.
- (9) Fichet G, Comoy E, Duval C, Antloga K, Dehen C, Charbonnier A, et al. *Novel methods for disinfection of prion-contaminated medical devices.* Lancet 2004; 364:521-526.
- (10) Glatzel M, Abela E, Maissen M, Aguzzi A. *Extraneural pathologic prion protein in sporadic Creutzfeldt-Jakob disease.* New Eng J Med 2004; 349:1812-1820.
- (11) Hannah EL, Belay ED, Gambetti P, Krause G, Parchi P, Capellari S, et al. *Creutzfeldt-Jakob disease after receipt of a previously unimplicated brand of dura mater graft.* Neurology 2001; 56:1080-1083.
- (12) Communicable Disease Network Australia. *Infection control guidelines for the prevention of transmission of infectious diseases in the health care setting.* 1-12. 2004.  
<http://www.health.gov.au/internet/wcms/publishing.nsf/content/icg-guidelines-index.htm>

- (13) Rohwer RG. *Assessment and management of risks from TSE contamination of blood and other tissues*. Presentation Public Health Agency of Canada International CJD Scientific Meeting. Ottawa. December 2004.
- (14) Rutala WA, Weber DJ. *Creutzfeldt-Jakob disease: recommendations for disinfection and sterilization*. Clin Infect Dis 2001; 32:1348-1356.
- (15) Somerville D. *Mechanisms of heat and chemical inactivation of TSE models*. Presentation Public Health Agency of Canada International CJD Scientific Meeting. Ottawa. December 2004.
- (16) UK CJD Incidents Panel. *Management of possible exposure to CJD through medical procedures: a consultation paper*. 1-62. 2001. London, Department of Health Publications. [http://www.hpa.org.uk/infections/topics\\_az/cjd/framework\\_Aug%202005.pdf](http://www.hpa.org.uk/infections/topics_az/cjd/framework_Aug%202005.pdf)
- (17) UK guidelines. *Distribution of TSE infectivity in human tissues and body fluids. Annex A.1, 1-2*. 2003. [http://www.advisorybodies.doh.gov.uk/acdp/tseguidance/tseguidance\\_annexa1.pdf](http://www.advisorybodies.doh.gov.uk/acdp/tseguidance/tseguidance_annexa1.pdf)
- (18) Weber DJ, Rutala WA. *Managing the risk of nosocomial transmission of prion diseases*. Curr Opin Infect Dis 2002; 15:421-425.
- (19) World Health Organization. *WHO infection control guidelines for transmissible spongiform encephalopathies: Report of the WHO consultation, Geneva, Switzerland, 23-26 March 1999*, 1-36. 2000. <http://www.who.int/csr/resources/publications/bse/whocdsraph2003.pdf>.
- (20) World Health Organization. *WHO guidelines on Transmissible Spongiform Encephalopathies in relation to biological and pharmaceutical products*. 2003. [http://www.who.int/bloodproducts/publications/en/WHO\\_TSE\\_2003.pdf](http://www.who.int/bloodproducts/publications/en/WHO_TSE_2003.pdf)
- (21) Will RG. *Iatrogenic Creutzfeldt-Jakob disease*. Presentation Public Health Agency of Canada International CJD Scientific Meeting. Ottawa. December 2004.
- (22) Wilson K. *Managing risk in the face of uncertainty: How useful is the precautionary principle*. Presentation Public Health Agency of Canada International CJD Scientific Meeting. Ottawa. December 2004.
- (23) Yan Z-X, Stitz L, Heeg P, Pfaff E, Roth K. *Infectivity of prion protein bound to stainless steel wires: A model for testing decontamination procedures for transmissible Spongiform Encephalopathies*. Infect Control Hosp Epidemiol 2006; 25:280-283.