

Infection Control Guidelines

Classic Creutzfeldt-Jakob Disease in Canada



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Health Canada

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Infection Control Guidelines

Classic Creutzfeldt-Jakob Disease in Canada

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Executive Summary

Problem

Transmissible spongiform encephalopathies (TSEs), also known as prion diseases, are fatal, degenerative brain diseases. The TSE agents are hardy, remain infectious for years in a dried state, and resist all routine sterilization and disinfection procedures commonly used in health care facilities. Although the incidence of Creutzfeldt-Jakob Disease (CJD) and other human TSEs is rare, there are increasing infection prevention and control concerns because of the unconventional nature of TSE agents and documented iatrogenic transmission between humans. The increasing availability of neurological and neurosurgical procedures has the potential to increase risks for iatrogenic transmission of the CJD agent if appropriate infection prevention and control measures are not taken.

Method

This guideline provides a framework within which institutions and agencies may develop policies and procedures to address their needs. The Health Canada infection control guidelines provide evidence-based recommendations. Where scientific evidence is lacking, the consensus of experts is used to formulate a recommendation. An overview of CJD and other human TSEs, including modes of transmission, is presented to provide a background for the recommendations that follow.

Risk Assessment

Tables and algorithms provide a risk assessment tool for decision making. Known iatrogenic sources of CJD are contaminated corneal and dura mater grafts, stereotactic electroencephalography electrodes and neurosurgical instruments, and human growth hormone and human pituitary gonadotropin. It is important to assess the patient and tissue risk for CJD to determine the infection prevention and control precautions necessary to prevent the transmission of CJD from one patient to other patients or care providers. Patients who are classified as high risk are those who are diagnosed with CJD and those who are suspected of having CJD on the basis of clinical signs and symptoms indicating progressive neurological disease. Patients at risk for CJD are those who have received human dura mater grafts, corneal grafts, and human pituitary hormones. Members of families with familial CJD, Gerstmann-Sträussler-Scheinker syndrome (GSS), and fatal familial insomnia (FFI) are also considered to be at risk for prion disease.

Assignment of different organs and tissues to categories of high and low infectivity is based upon the frequency with which infectivity has been detected under experimental conditions. High infectivity tissues are the brain, spinal cord, dura mater, pituitary, and eye (including optic nerve and retina). Low infectivity tissues are cerebrospinal fluid (CSF), kidney, liver, lung, lymph nodes, spleen, and placenta. There is no detected infectivity in adipose tissue, skin, adrenal gland, heart muscle, intestine, peripheral nerve, prostate, skeletal muscle, testis, thyroid gland, feces, milk, nasal mucus, saliva, serous exudate, sweat, tears, urine, blood, bone marrow, or semen. Although CSF is classified as low infectivity and is less

infectious than high infectivity tissues, it is felt that instruments contaminated by CSF should be handled in the same manner as those contacting high infectivity tissues in high risk and at risk patients.

Three algorithms have been developed to assist with decision making regarding the management of instruments and equipment used on a high risk or an at risk patient, depending on whether the instruments came into contact with tissues of high, low, or no detected infectivity.

Risk Management

The results of the risk assessment for CJD should be used to determine the appropriate infection prevention and control precautions required. The following table summarizes CJD decontamination processes that must be initiated for a high risk patient when exposure to high or low infectivity tissues is anticipated and for an at risk patient when exposure to high infectivity tissues (including CSF) is anticipated.

Table 6: CJD Decontamination Processes⁽¹⁾	
<i>These recommendations are based on the best available evidence at this time and are in decreasing order of effectiveness. They must be followed, without exception, when there is exposure to high and low infectivity tissues from a high risk patient and high infectivity tissues and CSF from an at risk patient (see Tables 2 and 3). NOTE: If the instrument or surface cannot be fully immersed or flooded with the chemical disinfectant, then the item must be incinerated.</i>	
1. Incineration: Use for all instruments, effluent materials, and solid waste.	
2. Instrument Decontamination for heat-resistant reusable instruments that an institution is unwilling or unable to incinerate.	
2.1	Cleaning: Removal of adherent particles through mechanical or manual cleaning must be completed prior to any chemical/sterilizer decontamination of instruments. Instruments and other materials to be decontaminated should be kept moist between the time of exposure to infectious materials and subsequent decontamination; then
2.2	Immerse instruments in 1N sodium hydroxide (NaOH) or sodium hypochlorite* solution for 1 hour; remove from chemical solution; rinse instruments well; then immerse instruments in water and place in a sterilizer selecting the liquid cycle and heat to 121° C for 1 hour; or
2.3	Immerse instruments in 1N NaOH or sodium hypochlorite* for 1 hour; remove instruments from chemical solution; thoroughly rinse instruments in water; then transfer to an open pan, place in prevacuum sterilizer, and heat to 134° C for 1 hour or to 121° C in a gravity displacement sterilizer for 1 hour.
3. Hard Surface Decontamination	
3.1	Remove visible soil.
3.2	Flood with 2N NaOH or undiluted sodium hypochlorite; let stand for 1 hour; then mop up and rinse with water; or

*20,000 ppm available chlorine

3.3	If surfaces cannot tolerate NaOH or undiluted sodium hypochlorite, thorough cleaning will remove most infectivity by dilution, and some additional benefit may be derived from the use of one or another of the partially effective methods listed in Table 5.
4. Chemical/Sterilizer Process for Dry Goods	
4.1	Small dry goods that can withstand either NaOH or sodium hypochlorite should first be immersed in one or the other solution (as described in 2.3 above) and then heated in a prevacuum steam sterilizer to 134° C for 1 hour.
4.2	Bulky dry goods or dry goods of any size that cannot withstand exposure to NaOH or sodium hypochlorite should be heated in a prevacuum steam sterilizer to 134° C for 1 hour.

Recommendations

The recommendations in this guideline represent a conservative approach to the management of classic CJD in health care and public service settings in Canada. Recommendations on the management of a high risk or an at risk patient for CJD in the health care setting include medical and surgical procedures and, for high risk patients, precautions during childbirth and dentistry. The recommendations elaborate on the CJD decontamination process for instruments and equipment. The use of single-use instruments and equipment is recommended wherever possible for high infectivity tissue and CSF when CJD is a concern. Other recommendations deal with laboratory specimen collection and handling, and pathology; autopsy and mortuary procedures; and issues related to occupational safety.

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Health Canada's Infection Control Guidelines Program

The primary objective in developing guidelines at the national level is to help health care professionals improve the quality of health care. Guidelines for infection prevention and control are needed to assist in the development of policies, procedures, and evaluative mechanisms to ensure an optimal level of care. Guidelines, by definition, are directing principles and indications or outlines of policy or conduct, which should not be regarded as rigid standards. Guidelines facilitate the setting of standards but respect the autonomy of each institution and recognize the governing body's authority and responsibility in ensuring that patient or resident care provided by the institution is of high quality.

Wherever possible, these guidelines have been based on research findings. There are, however, some areas in which the published research is very limited; consequently, the consensus of experts in the field has been used to provide guidelines specific to conventional practice in these situations. Health Canada invited experts working in public health, prion research, infection control, infectious diseases, instrument reprocessing, sterilizer manufacturing, occupational health, neurosurgery, ophthalmic surgery, and operating room management to a consensus meeting on January 11, 2002, to finalize the recommendations in this guideline.

The information in the guideline was current at the time of publication; it should be emphasized that areas of knowledge and aspects of medical technology advance with time. Health care professionals are encouraged to contact Health Canada for updated information. Both encouragement of research and frequent revision and updating to keep pace with advances in the field are necessary if guidelines are to achieve the purpose for which they have been developed.

The Health Canada Infection Control Guidelines Steering Committee acknowledges, with sincere appreciation, the many practising health professionals and others who contributed advice and information to this endeavour. Health Canada is especially appreciative of the time and expertise contributed by the Working Group.

This guideline is part of a series that has been developed over a period of years under the guidance of the Steering Committee on Infection Control Guidelines. *Classic Creutzfeldt-Jakob Disease in Canada: An infection control guideline* represents an overview of Creutzfeldt-Jakob Disease (CJD) and recommendations to assist in the prevention of the exposure to and transmission of CJD and other human transmissible spongiform encephalopathies to patients and workers in health care facilities and to public service workers.

This document is part of the Health Canada series of Infection Control Guidelines and is intended to be used with the other infection control guidelines. Others in the series include the following:

- Prevention and Control of Occupational Infections in Health Care (2002)
- Routine Practices and Additional Precautions for Preventing Transmission of Infection in Health Care (1999)
- Infection Prevention and Control Practices for Personal Services: Tattooing, Ear/Body Piercing and Electrolysis (1999)
- Hand Washing, Cleaning, Disinfection and Sterilization in Health Care (1998)
- Preventing the Spread of Vancomycin-Related Enterococci (VRE) in Canada (1997)
- Preventing Infections Associated with Foot Care by Health Care Providers (1997)
- Preventing Infections Associated with Indwelling Intravascular Access Devices (1997)
- Preventing the Transmission of Bloodborne Pathogens in Health Care and Public Services Settings (1997)
- Canadian Contingency Plan for Viral Hemorrhagic Fevers and Other Related Diseases (1997)
- Preventing the Transmission of Tuberculosis in Canadian Health Care Facilities and Other Institutional Settings (1996)
- Long Term Care Facilities (1994)
- Prevention of Nosocomial Pneumonia (1990) (under revision)
- Antimicrobial Utilization in Health Care Facilities (1990)
- Prevention of Surgical Wound Infections (1990)
- Prevention of Urinary Tract Infections (1990)
- Organization of Infection Control Programs in Health Care Facilities (1990)
- Perinatal Care (1988)

Another publication of the Nosocomial and Occupational Infections section of the Health Care Acquired Infections Division that complements the Infection Control Guidelines series is *Construction-related Nosocomial Infections in Patients in Health Care Facilities: Decreasing the Risk of **Aspergillus**, **Legionella** and Other Infections* CCDR 2001 27S2.

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Introduction

A. How to Use this Document

This document presents an overview of CJD and other human transmissible spongiform encephalopathies (TSEs) (see Part A), an approach to the assessment and management of CJD risk in the clinical setting (see Part B), and recommendations to assist in the prevention of exposure to and transmission of CJD and other human TSEs (e.g. Gerstmann-Sträussler-Scheinker and fatal familial insomnia) to patients and workers in health care facilities and to public service workers (see Part C). Readers who are unfamiliar with CJD should use this document and consult with experts in the field. The World Health Organization (WHO) *Infection Control Guidelines for Transmissible Spongiform Encephalopathies*⁽¹⁾ has been referenced extensively throughout this document. The information from the WHO and current research has been summarized and, in some areas, adapted for the Canadian context, and a classification system has been used to assist the reader in weighing the extent and strength of evidence behind each recommendation (see Appendix I).

Several countries have prepared guidelines for infection prevention and control related to CJD. However, some recommendations vary from guideline to guideline because specific scientific evidence to support one practice is often lacking. Additionally, there are issues on which consultants cannot agree or for which there is insufficient evidence upon which to base a recommendation. Recommendations are based on an interpretation of the limited amount of information available at the time they are prepared. Consequently, as information becomes available it will be reviewed and changes made where appropriate. The recommendations outlined in this document represent a conservative approach to the management of classic CJD in health care facilities and public service settings in Canada.

There is no reason to deny necessary procedures to a patient with CJD or another human TSE. Careful planning is required when caring for these patients to avoid unnecessary exposures that could necessitate destruction of expensive equipment. Special decontamination procedures are required for devices that come into contact with certain tissues of patients at high risk or at risk for classic CJD.

These guidelines provide a framework within which institutions and agencies may develop policies and procedures to address their needs. The overview of prion diseases and issues relevant to infection prevention is presented to provide a background for the recommendations that follow. Table 2 (Patient Risk for CJD), Table 3 (Tissue Risk for CJD) and Table 4 (Infection Prevention and Control Management Based on Risk Assessment for CJD) should help infection prevention and control personnel in problem solving for situations not specifically addressed in these guidelines. Appendix V provides an example of a hospital tool for risk assessment and management based on the Health Canada guideline.

B. Glossary and Abbreviations (See Appendix II for CJD Surveillance Definitions)

<i>At risk patient</i>	... Asymptomatic patient who has identified risk factors for developing CJD or another human TSE (e.g., GSS, FFI), excluding variant CJD (see Table 2)
<i>Autoclave</i>	... Gravity displacement steam autoclave: Sterilization system in which incoming steam displaces residual air through a port or drain that is situated at the lowest point of the sterilizer chamber ⁽²⁾ . A gravity displacement autoclave is designed for general decontamination and sterilization of solutions and instruments. ... <i>Prevacuum (porous load) steam autoclave</i> : Sterilization system in which air is exhausted by vacuum and replaced by steam. A <i>prevacuum (porous load) system autoclave</i> is best suited for sterilization of clean instruments, gowns, drapes, towelling and other dry materials required for surgery. It is not suited for sterilization of liquids.
<i>BSE</i>	... Bovine spongiform encephalopathy – a transmissible spongiform encephalopathy affecting cattle, more commonly known as mad cow disease.
<i>CJD</i>	... All forms of classic Creutzfeldt-Jakob disease (sporadic, familial and iatrogenic), excluding variant CJD
<i>CJD precautions</i>	... A new category within the Additional Precautions for Preventing the Transmission of Infection in Health Care ⁽³⁾ . CJD precautions are based on patient and tissue risks for CJD and are necessary only for well-defined situations.
<i>CJD-SS</i>	... Creutzfeldt-Jakob Disease Surveillance System (Canada)
<i>CNS</i>	... Central nervous system
<i>CSF</i>	... Cerebrospinal fluid
<i>Decontamination</i>	... The process of cleaning, followed by the inactivation of pathogenic microorganisms, in order to render an object safe for handling ⁽⁴⁾

<i>Dementia</i>	... An organic mental disorder characterized by a general loss of intellectual abilities involving impairment of memory, judgement, and abstract thinking as well as changes in personality
<i>Device</i>	... A medical device, instrument or equipment used in health care
<i>Disinfection</i>	... The inactivation of disease-producing microorganisms. Disinfection does not destroy bacterial spores. Disinfection usually involves chemicals, heat or ultraviolet light.
<i>Dura mater</i>	... The outermost of the three membranes covering the brain and spinal cord
<i>Familial CJD</i>	... A form of CJD occurring in families, associated with mutations in the prion protein (PrP) gene
<i>FFI</i>	... Fatal familial insomnia: a rare type of inherited human TSE associated with a mutation in the prion protein (PrP) gene
<i>Gonadotropin</i>	... Any hormone having a stimulating effect on the gonads, such as those secreted by the pituitary gland
<i>GSS</i>	... Gerstmann-Sträussler-Scheinker disease: a rare type of inherited human TSE associated with a mutation in the prion protein (PrP) gene
<i>HCW(s)</i>	... Health care worker(s)
<i>hGH</i>	... Human growth hormone
<i>High risk patient</i>	... Patient with a clinical diagnosis of CJD or another human TSE (e.g., GSS, FFI) or suspected of having one of these diseases (see Table 2), excluding variant CJD.
<i>Iatrogenic CJD</i>	... A rare form of CJD introduced accidentally in a patient as the result of a medical procedure in which there has been exposure to infectious tissue.

<i>Kuru</i>	... A type of human TSE, which occurred in epidemic form in the Fore people of Papua New Guinea in the 1950s and was linked to ritualistic cannibalism.
<i>MRI</i>	... Magnetic resonance imaging is a type of diagnostic radiography using electromagnetic energy.
<i>Pituitary hormone</i>	... Hormone derived from the pituitary gland, such as growth hormone (GH) and follicular stimulating hormone (FSH)
<i>Prion protein</i>	... A protein that is present in many organs and tissues, including the brain, spinal cord and eye of healthy humans and animals. The TSE agent is believed to be an abnormal form of a prion protein that causes surrounding prion proteins to change their conformation.
<i>PrP</i>	... Prion protein (PrP ^c is the normal endogenous cellular protein, PrP ^{Sc} is the pathogenic abnormal conformation)
<i>Sporadic CJD</i>	... The most common form of CJD, which appears to occur spontaneously with no identifiable cause.
<i>TSE</i>	... Transmissible spongiform encephalopathy – a fatal degenerative brain disease that occurs in humans and certain animal species, also known as prion disease.
<i>Variant CJD</i>	... Variant Creutzfeldt-Jakob disease

C. The Purpose of the Infection Prevention and Control Guideline for CJD and Other Human TSEs

TSEs, also known as prion diseases, are fatal degenerative brain diseases that occur in human and certain animal species. They are characterized pathologically by microscopic vacuoles (spongiform change) and the deposition of amyloid (prion) protein in the grey matter of the brain⁽¹⁾. There is no known treatment for these diseases, and the outcome is uniformly fatal. The TSE agents are hardy, remain infectious for years in a dried state, and resist all routine sterilization and disinfection procedures commonly used by health care facilities^(1,5,6). All forms of TSE are experimentally transmissible^(1,7).

In animals, examples of these diseases include scrapie in sheep and goats, bovine spongiform encephalopathy (BSE) in cattle, chronic wasting disease in deer and elk, and transmissible mink encephalopathy. In humans, four diseases were originally identified as TSEs: Creutzfeldt-Jakob disease (CJD), Gerstmann-Sträussler-Scheinker syndrome (GSS), fatal familial insomnia (FFI), and kuru^(7,8). Two new forms of human TSEs have recently been identified: variant CJD and sporadic fatal insomnia^(9,10). Appendix II provides the surveillance definitions for classic CJD (sporadic, iatrogenic and familial), GSS, FFI, and kuru.

Although the incidence of CJD and other human TSEs is rare, there are increasing infection prevention and control concerns with regard to these diseases because of the unconventional nature of TSE agents and the documented iatrogenic transmission between humans, together with the increasing media attention following the BSE epidemic in Europe and the appearance of variant CJD in Europe in the mid-1990s. The increasing availability of neurological and neurosurgical procedures developed over the past decades have improved patient care, but may have the potential to increase the risk of iatrogenic transmission of the CJD agent if appropriate infection prevention and control measures are not taken.

In response to increased awareness and heightened surveillance of CJD in Canada, the Centre for Infectious Disease Prevention and Control in the Population and Public Health Branch, Health Canada, formed a multi-disciplinary committee to examine the issues in relation to CJD and infection prevention and control practices. Recent findings on the limitations of decontamination procedures and new information on CJD prompted the committee to recommend a thorough review of the literature, existing guidelines⁽¹¹⁻¹³⁾ and established protocols.

This guideline was based on the results of a review of the research evidence and in consultation with experts. It is acknowledged that information is limited at this time and that decontamination recommendations are based primarily on animal models. The purpose of this document is to provide guidance for precautions to prevent or minimize exposure of both patients and workers to CJD agents in health care and public service settings.

In mid-2002, a case of variant CJD was found in Canada in an individual who had resided for some time in the U.K. Epidemiologists have concluded that this case was acquired during residency in the U.K. Current information on variant CJD indicates that lymphoreticular tissues are involved and that the infectivity is different from that of other forms of CJD. Patients with variant CJD might therefore pose a greater risk of transmitting iatrogenic infections than those with sporadic CJD⁽¹⁾. Consequently, risk tissues and decontamination procedures will need to be redefined for variant CJD as further research findings emerge⁽¹²⁾. A brief description of variant CJD is outlined later in this document, but infection prevention and control measures to prevent or minimize exposure to variant CJD **will not** be addressed. If a case of variant CJD is clinically suspected or diagnosed, the practitioner should consult with Nosocomial and Occupational Infections, Health Care Acquired Infections Division, Centre for Infectious Disease Prevention and Control, Population and Public Health Branch, Health Canada, for specific infection prevention and control measures.

D. Goals of the Guideline

1. To aid in the identification of individuals and situations that pose a risk for iatrogenic transmission of CJD
2. To provide recommendations to minimize the risk of transmission of CJD from patient to patient
3. To provide guidance for the protection of workers in health care facilities and public service settings
4. To provide a working document as a decision-making tool for situations not discussed or anticipated by the guideline.

PART A.

OVERVIEW OF CJD AND OTHER HUMAN TSEs

Background Information

A. Etiology

The agent causing CJD and other human TSEs has not yet been definitively identified. It was originally thought that the infectious agent was a slow virus or virus-like particle^(7,13). However, increasing evidence suggests that unconventional agents, termed prion proteins (PrP), are central in the etiology of these diseases^(14,15). The prion is theorized to contain only protein, has no DNA or RNA, and replicates by converting the structure of the normal cellular prion protein into an abnormal one^(8,14,16,17).

B. Clinical Features of CJD

CJD was first described in the 1920s and is by far the most common TSE. The incubation period can extend up to 30 years^(18,19). Although there is a wide spectrum of clinical features, the typical clinical presentation is a progressive dementia that soon becomes associated with progressive unsteadiness and clumsiness (cerebellar ataxia), visual deterioration, muscle twitching (myoclonus), and a variety of other neurological symptoms and signs. The affected person is usually mute and immobile in the terminal stages, and in most cases death occurs within a few months of onset of symptoms (mean 5 months)^(8,17,20,21). CJD is invariably fatal, and there is no treatment⁽¹⁾.

Three forms of classic CJD are recognized: sporadic, familial, and iatrogenic. The sporadic form, which accounts for 85% to 90% of all cases^(1,7,18,22), occurs apparently spontaneously in the general population with no identifiable cause. Eighty percent of sporadic cases are diagnosed in people between 50 and 70 years of age⁽⁸⁾ (average 60 years^(18,23)). The familial form accounts for approximately 10% to 15% of all cases^(8,22) and results from mutations in the coding sequence of the (PrP) gene⁽²⁴⁾. GSS and FFI are exceptionally rare human TSEs and also appear to be inherited.

Documented iatrogenic CJD is very rare, accounting for less than 1% of all known cases. It is the result of person-to-person transmission arising from invasive medical interventions in which the source patient had CJD^(1,18,24,25). Attempts have been made to distinguish between the clinical characteristics of patients with iatrogenic CJD and those with sporadic CJD, in order to differentiate the origin of the infection. In one study (n = 20), all those with iatrogenic CJD after pituitary hormone therapy presented with cerebellar signs, as compared with 33% of sporadic CJD cases⁽²⁶⁾. A recent review of all cases of iatrogenic CJD up to July 2000 indicated that cerebellar onset predominates in recipients of pituitary hormone and dura mater grafts⁽¹⁹⁾. Although clinical presentation can contribute to increased suspicion

that a case is iatrogenic, it cannot definitively distinguish iatrogenic from sporadic CJD.

C. Diagnosis of CJD

CJD is not readily diagnosed, as there is no diagnostic test available that uses easily accessible biological tissue, nor are there ready means to determine whether there has been exposure to the infectious agent. The diagnosis of CJD is currently based on clinical features with the support of investigations, and confirmed by postmortem neuropathological examination. Characteristic electroencephalogram (EEG) changes, such as the presence of triphasic periodic complexes, are often of assistance in making the diagnosis⁽²²⁾. Repeated EEGs may be required before the diagnosis can be made. The typical EEG appearance has only rarely been seen in iatrogenic disease related to growth hormone.

Imaging techniques are useful in excluding other causes of subacute dementia. A lumbar puncture is often done to exclude other disease processes. The cerebrospinal fluid (CSF) does not show an increase in cells and has a normal or mildly elevated protein content⁽⁸⁾. A diagnostic test for detection of 14-3-3 protein in CSF has a high sensitivity and specificity for diagnosis of sporadic CJD during the clinical illness⁽²²⁾. The WHO Consultation on Global Surveillance, Diagnosis, and Therapy for Human TSEs, held in Geneva in February 1998, discouraged the use of cerebral biopsy in living patients who are suspected of having CJD, except to make an alternative diagnosis of a treatable disease⁽²⁰⁾. A definite diagnosis of CJD or other human TSE (GSS, FFI, or variant CJD) is established only by neuropathological examination⁽²²⁾.

D. Epidemiology and Surveillance

CJD has been identified in all developed countries, and the incidence worldwide is between 0.5 and 1 case per million population per year^(18,20,24). The Canadian epidemiologic pattern is similar to that found elsewhere^(17,27).

The Centre for Infectious Disease Prevention and Control, in the Population and Public Health Branch, is coordinating an intensive active surveillance program – the Creutzfeldt-Jakob Disease Surveillance System (CJD-SS) – to determine the current incidence of CJD and other human TSEs in Canada⁽¹⁷⁾. As a member of an international project, CJD-SS also conducts surveillance for variant CJD.

The CJD-SS was launched in 1998. Cases are reported to the study coordinator by neurologists, geriatricians, neurosurgeons, neuropathologists, infection control practitioners, and infectious disease physicians, who are asked to seek consent from the patient and family for participation. If there is agreement to participate, data collection includes 1) an interview to obtain demographic information, family history, and exposure history; 2) a review of medical records for evidence of exposure to blood and blood products; 3) the testing of CSF samples by 14-3-3 assay; and 4) the sequencing of the protein coding sequence of the PrP gene using DNA extracted from blood. The CJD-SS can help in the arrangements for autopsy and also provides neuropathology and immunohistochemistry services, on request, for

final case identification. So far, the CJD-SS has identified three cases of iatrogenic transmission of CJD following dura mater graft⁽²⁸⁾.

Appendix II provides the surveillance definitions used in Canada for sporadic CJD (confirmed, probable and possible), iatrogenic CJD, familial CJD, GSS, FFI and kuru.

It is expected that case notification will come primarily from neurologists, geriatricians, neurosurgeons, neuropathologists, infection control practitioners, and infection disease physicians. However, any practitioner aware of a suspected or diagnosed case of CJD is asked to contact the surveillance system for non-nominal notification at Health Canada's toll free number: 1-888-489-2999.

E. Variant CJD

Variant CJD was first recognized in the United Kingdom (U.K.) in 1996⁽¹⁰⁾. As of April 2002, the worldwide reported definite and probable cases of variant CJD include 117 cases in the U.K.^(29,30), 6 in France⁽³¹⁻³⁴⁾, 1 in the Republic of Ireland⁽³⁵⁾, 1 in Hong Kong⁽³⁶⁾, and 1 in Italy⁽³⁷⁾. Epidemiologic and scientific evidence indicates that the agent responsible for BSE in cattle is the same agent responsible for variant CJD^(8,38,39). The outbreak of variant CJD most probably is the result of consumption of beef or beef products from BSE-infected cattle⁽⁴⁰⁾.

Variant CJD differs in several ways from sporadic CJD. The individuals affected are younger, with a median age at death of 29 years⁽⁴¹⁾. The presenting features are often psychiatric disturbances (e.g., anxiety, depression, withdrawal) and sensory symptoms (e.g., persistent paresthesia or dysesthesia), followed by other neurological symptoms and progressive cognitive impairment⁽⁴¹⁾. Variant CJD has a marginally longer duration of illness (median 14 months, range 8-38 months) than does classic CJD⁽⁴¹⁾. The electroencephalograms, though not normal, do not show the typical triphasic wave complexes found in sporadic CJD⁽⁴¹⁾. Magnetic resonance imaging (MRI) brain scans show bilateral pulvinar high signal in over 70% of cases^(41,42). The neuropathology in variant CJD differs in key respects from other human TSEs. It is characterized by the presence of prominent spongiform change and extensive PrP deposition with florid plaques throughout the cerebrum and cerebellum^(8,14,20).

The abnormal PrP molecular strain type (type 4) is unique and distinctive in variant CJD patients^(39,43). It has been detected in lymphoreticular tissues of patients who have died from variant CJD but not in such tissues of patients with other forms of CJD⁽⁴³⁾. Variant CJD can be diagnosed in the appropriate clinical context by tonsil biopsy^(29,43,44).

In mid-2002, one case of variant CJD was identified in Canada by the CJD-SS. On April 18, 2002, the United States reported a likely case of variant CJD⁽⁴⁵⁾. The one case of BSE that was reported in Canada occurred in 1993 in a cow that had been imported from the U.K. in 1987. Since 1993, no other cases of BSE have been reported, and BSE is not known to exist in Canada⁽⁴⁶⁾.

Transmission of CJD

A. Overview

CJD was recognized as a transmissible disease in the mid-1960s, when kuru (a human TSE of the Fore people of Papua New Guinea) was shown to be transmissible and when CJD was experimentally transmitted to chimpanzees^(21,47). CJD and other human TSEs are not known to spread by contact from person to person or by the airborne/respiratory route⁽¹⁾. However, transmission can occur during invasive medical interventions^(1,19). Worldwide, as of July 2000, there have been 267 documented transmissions of CJD between humans since the first report in 1974, and new cases continue to appear⁽¹⁹⁾. There are three circumstances in which transmission of CJD between humans has been demonstrated: transplantation of central nervous system (CNS) tissue, use of contaminated instruments used during invasive neurological or neurosurgical procedures, and peripheral administration of human pituitary extracts^(19,24). The known iatrogenic causes of CJD are summarized in Table 1.

Normal social and clinical contact, and non-invasive clinical investigations (e.g. x-ray imaging procedures) with CJD patients do not present a risk to health care workers, relatives, or the community⁽¹⁾.

Table 1: Known Iatrogenic Causes of CJD (July 2000)		
Mode of Infection	Number of Patients	Examples
Tissue Transplantation Corneal graft Dura mater graft	3 ^(8,19,48) 114 ⁽¹⁹⁾	Person-to-person transplantation of corneal ^(49,50) or dura mater grafts obtained from infected individuals ⁽⁵¹⁻⁵⁴⁾ , including two cases after embolization procedures with dura mater ^(55,56)
Instrumentation Neurosurgery Stereotactic EEG	5 ^(19,48,57) 2 ^(19,48,58,59)	Use of neurosurgical instruments or stereotactic depth electrodes that were contaminated by brain contact during prior use on infected individuals and were inadequately cleaned and/or sterilized ^(11,19,23,57,59,60)
Tissue Extract Transfer Growth hormone Gonadotropin	139 ^(8,19) 4 ⁽¹⁹⁾	Peripheral injections of human cadaveric growth hormone ^(61,62) or human pituitary gonadotrophin ⁽⁶³⁾ for treatment of endocrine disorders

Note: Person-to-person transmission via any other route has not been demonstrated.

From observation of known human iatrogenic cases, when CJD exposure is central (e.g. there is direct application of the CJD agent into the brain during neurosurgery), the incubation periods are relatively short, ranging from 12 to 28 months (median 17 months)^(19,26). As the inoculation site moves further away from the brain to other tissues, the incubation period is extended. For example, incubation periods have ranged from 1.5 to 18 years (median 6 years) following exposure to contaminated dura mater⁽¹⁹⁾. Transmission from exposure through a peripheral route (as with human growth hormone injections) is associated with an incubation period ranging from 5 to 30 years (median 12 years)⁽¹⁹⁾.

B. Transplantation of Central Nervous System Tissue

CNS tissue transplants include tissues such as corneal and dura mater grafts. In 1974, the first case of iatrogenic CJD was reported in the recipient of a corneal graft from a donor who had died from CJD, which had not been diagnosed before death^(19,49). Experimental evidence has demonstrated that corneas of infected animals can transmit CJD and that the causative agent spreads along visual pathways^(24,64). To date, three cases of corneal graft-related CJD have been reported, none of them in Canada^(8,19,48,50).

The first case of CJD in the recipient of a cadaveric-derived dura mater transplant was reported in 1987⁽⁵³⁾. As of July 2000, 114 cases have been reported worldwide⁽¹⁹⁾, four such cases occurring in Canada^(19,28,65). Nearly all the contaminated dura mater tissue was produced before 1987 by a single manufacturer, whose manufacturing processes were inadequate to inactivate the CJD agent^(24,66-68). This, combined with inadequate donor screening and pooling of dura mater⁽²⁴⁾, led to the relatively large number of cases. Although the tissue preparation process was modified in 1987, cases from earlier dura mater grafting continue to appear because of the long incubation period^(19,24,68). Stringent donor screening and processing of dura mater may not completely eliminate the potential for an infectious graft⁽⁶⁷⁻⁶⁹⁾. Health Canada no longer permits the sale of human dura mater in Canada^(65,70). Hospitals that perform neurosurgery use synthetic materials, non-dural human tissues, and animal tissues for dural repair⁽⁷¹⁾.

C. Instruments Used During Invasive Neurological and Neurosurgical Procedures

In 1977, Bernoulli et al. published the first reports of transmission of CJD as a result of direct brain instrumentation involving silver electrodes previously used in the brain of a person with CJD⁽⁵⁹⁾. The electrodes had been reprocessed with 70% alcohol and formaldehyde vapour⁽⁵⁹⁾. Support for the conclusion that transmission was iatrogenic followed from studies in which the same deep brain (intracerebral) recording electrodes were shown to transmit disease to experimental primates⁽²³⁾. Retrospective studies have revealed four to six other cases of probable iatrogenic transmission occurring as a consequence of neurosurgical procedures^(1,19,48,57,58). It is presumed that, in these cases, the routine sterilization procedures used for the instruments were insufficient to eliminate infectivity. Some of the transmissions associated with reprocessed instruments occurred several weeks after the

instruments had initially been exposed to tissues from a CJD patient. No such cases have been reported in Canada.

The rate of transmission from a single contaminated instrument is unknown, although it is certainly not 100%^(21,57), despite the direct route of application. Even given these few cases of transmission occurring during neurosurgical procedures, it is probable that the current risk of CJD transmission by means of neurosurgical instruments is extremely remote. There is no human evidence of CJD transmission from patients who are asymptomatic for CJD and not at increased risk of developing the disease^(19,72)(see Table 2). Therefore, such patients do not need CJD-specific infection control precautions⁽⁷²⁾.

The incidence of CJD in Canada does not justify the universal use of anything other than routine sterilization procedures for neurosurgical patients, and CJD precautions should be initiated only in the presence of a patient and tissue risk for CJD. However, consideration may be given, where feasible, to segregating neurosurgical instruments from all other instruments during routine cleaning in order to minimize exposure to potentially CJD contaminated instruments.

D. Peripheral Administration of Human Pituitary Extracts

The largest outbreak of CJD is ongoing among recipients of human growth hormone (hGH). By 1985, it was recognized that injected, human cadaver-extracted pituitary growth hormone could transmit CJD to humans⁽⁶²⁾. Shortly thereafter, it was also recognized that human pituitary gonadotropin administered by injection could transmit CJD between humans⁽⁶³⁾. As of July 2000, 143 cases of CJD worldwide have been related to growth hormone and gonadotropic hormone of human origin⁽¹⁹⁾. The use of cadaver-extracted growth hormone has now been replaced by recombinant growth hormone, which does not pose a risk of CJD. However, because of the long incubation period of CJD it is likely that further cases due to exposure to hGH will appear in years to come⁽²⁶⁾. In Canada, where human growth hormone was used from 1965 until April 1985, surveillance for hGH-associated CJD has not identified any such cases^(27,73).

E. Blood

There is no evidence to support human transmissibility of CJD through blood or blood products, and this remains only a theoretical risk. The results of experimental studies investigating the infectivity of blood are conflicting, and even when infectivity has been detected it is present in very low amounts⁽¹⁾. Some experimental evidence suggests that certain blood components can occasionally transmit the CJD agent⁽¹²⁾. However, because these experiments have involved direct injection of blood into the brains of animals (e.g. mice), extrapolation of the findings to blood transfusion as a route of infection is problematic^(74,75). A comprehensive review of three decades of research with non-human primates by the National Institutes of Health indicated that the blood of humans with CJD injected either peripherally or centrally into primates did not transmit CJD⁽²¹⁾.

Despite the possible presence of low levels of the CJD agent in the blood of some donors during the symptomatic stage of CJD, no case of transmission in humans through blood or from blood donors has ever been identified⁽⁷⁵⁾. Surveillance systems have found cases of CJD among people who have received blood transfusions, but none has been directly linked to the transfusion itself⁽²⁴⁾.

At present, no validated screening assays are available to detect PrP in asymptomatic people. However, to ensure that there is a large margin of safety in Canada's blood supply, Health Canada has recommended that people who are at risk of developing CJD and blood relatives (parent, child, sibling) of people with familial CJD be deferred from donating blood or blood products⁽⁷⁶⁾.

There has never been a reported case of transmission of variant CJD through blood or blood products^(77,78). However, experimental studies and the unique biological characteristics of variant CJD suggest that an increased theoretical risk of transmission of the infective agent in lymphocytes may exist with blood transfusion^(14,78).

Since August 1999, in accordance with the precautionary principle⁽⁷⁹⁾, Health Canada has issued several directives requiring the deferral of blood donors. In the Health Canada policy of August 2001, *Donor Exclusion to Address Theoretical Risk of Transmission of vCJD through the Blood Supply*, there are three categories of exclusion: people who have spent a cumulative period of 3 months or more in the United Kingdom or France between the years 1980 and 1996; those who have spent a cumulative period of 5 years or more in Western Europe from 1980 to the present; and those who received a transfusion of whole blood or blood components in the U.K. from 1980 to the present^(77,80,81). The exclusion dates coincide with the time when potential exposure to beef contaminated with BSE was considered to be at its peak.

F. Occupational Exposure

There is no epidemiologic evidence to indicate that health care workers (HCWs) are at increased occupational risk for CJD^(72,82). In most countries, measures to protect HCWs from occupational exposure to CJD have either been implemented or introduced only recently. Approximately 40 reports of CJD in individuals whose work puts them in contact with patients with diagnosed or suspected CJD have been found in the literature (e.g. case reports, population-based case series and case control studies)^(83,84). These individuals include physicians, neurosurgeons, an orthopedic surgeon, a pathologist, nurses, dentists, a dental surgeon, and histology technicians. Although a link with their occupation is suggested, none of these cases had been confirmed as having been acquired through occupational exposure⁽⁶⁰⁾. An analysis of four case control studies did not identify HCWs as being at higher risk for CJD⁽⁸³⁾. In keeping with the general population risk, a certain number of HCWs could be expected to develop CJD unrelated to their occupation. The incidence of CJD is not higher in this occupational group than in the general population.

However, in view of the documented transmissions of CJD between humans, it is prudent to take a precautionary approach and assume a theoretical risk of transmission related to occupational exposure. In this context, the highest potential risk is from exposure to high infectivity tissues from a high risk patient (see Tables 2 and 3) through needlestick injuries with inoculation⁽¹⁾. Therefore, percutaneous exposures to high or low infectivity tissues from a high risk patient and high infectivity tissues (including CSF) from an at risk patient must be avoided (see Tables 2 and 3). For recommendations on reducing the risk of percutaneous injuries, refer to Health Canada's *Infection Control Guidelines Preventing the Transmission of Bloodborne Pathogens in Health Care and Public Services Settings*⁽⁸⁵⁾.

PART B. ASSESSMENT AND MANAGEMENT OF CJD RISK IN CLINICAL PRACTICE

Risk Assessment for CJD

When considering infection prevention and control precautions to prevent the transmission of CJD from a patient to other individuals (patients, health care workers, or other care providers), it is important to assess the patient and tissue risk for CJD. The risk assessment is dependent upon two factors: 1) the probability that a patient has or will develop CJD, and 2) the level of infectivity in the patient tissues. Using these two factors, it is possible to make a decision as to what CJD precautions may be required.

A. Patient Risk Assessment for CJD

Patients with diagnosed or suspected CJD (as well as GSS and FFI) pose the highest risk for transmission of prion disease⁽¹⁾ (see Table 2).

Table 2: Patient Risk for CJD ⁽¹⁾	
High Risk Patient	At Risk Patient*
<p>Diagnosed CJD**</p> <p>Suspected CJD:** undiagnosed, unusual, progressive neurological disease consistent with CJD** (e.g. dementia, myoclonus, ataxia, etc.)</p>	<p>Only under conditions in which there could be exposure to their high infectivity tissues, including CSF (see Table 3):</p> <p style="padding-left: 40px;">Recipients of human dura mater grafts,[†] corneal grafts,[§] and human pituitary hormones</p> <p style="padding-left: 40px;">Members of families with familial CJD, GSS and FFI[¶]</p>

* The incidence of CJD in Canada does not justify classifying people who have undergone neurosurgical procedures as at risk patients.

** All forms of classic CJD (sporadic, familial and iatrogenic), GSS and FFI, excluding variant CJD

† Recipients of dura mater grafts are frequently unaware of having received the graft.

§ Corneal grafts originating in a jurisdiction that requires the graft donor to be evaluated for neurological disease are not considered a risk for CJD⁽⁷⁶⁾.

¶ Familial CJD, GSS and FFI can be determined by genetic testing or may be indicated by the occurrence of two or more cases of CJD, GSS or FFI in the family (parent, child, sibling)⁽⁷⁶⁾.

At the WHO Consultation on Infection Control Guidelines for Transmissible Spongiform Encephalopathies, held in March 1999, it was felt that at risk patients (recipients of human dura mater grafts, corneal grafts and human pituitary hormones, and people who have undergone neurosurgical procedures) present a risk of transmitting the CJD agent only under conditions in which there could be exposure to their high infectivity tissues (see Table 3). The WHO consultants

considered that appropriate control measures have immensely reduced or eliminated exposure to contaminated dura mater and pituitary hormones, and noted that there have been only three reports of CJD transmission through corneal transplantation and seven reports (all before 1980) of transmission via instruments used during invasive neurological or neurosurgical procedures. It was also recognized that recipients of dura mater are largely unaware that they are recipients, making identification of many of them unlikely⁽¹⁾.

Currently, world experts have not reached consensus as to whether an asymptomatic person at risk for familial CJD, GSS or FFI should be classified as at risk when the need for CJD precautions is being determined. As scientific resolution is impossible at this time because of lack of precise information about tissue infectivity during the pre-clinical phase of human TSEs, a conservative approach to infection prevention and control has been adopted in these guidelines.

B. Tissue Risk Assessment for CJD

Information describing the degree of infectivity for CJD is derived largely from experiments with animal TSEs (e.g. BSE and scrapie), which have established that different organs and tissues have different infectivity levels in different species^(21,86). In general, the higher the concentration of the CJD agent in the tissues of origin, the greater the assumed risk of transmission and the shorter the incubation period before the onset of symptoms.

The CJD agent is highly concentrated in the brain and central nervous system^(1,21,87). Investigations of iatrogenic CJD cases resulting from injections of human growth hormone and gonadotropin extracted from pituitary glands have demonstrated that a concentrated dose, even delivered by a peripheral route, can result in transmission of CJD^(62,63).

Table 3 categorizes the tissue risk for transmission of CJD by the level of infectivity found in different organs and tissues (high, low, and no detected infectivity). It is a guide to predicted infectivity of body tissues and fluids. Assignment of different organs and tissues to categories of *high* and *low infectivity* is based chiefly on the frequency with which infectivity has been detected rather than on quantitative assays of the level of infectivity, for which data are incomplete. Experimental data are derived from primates inoculated with tissues from human cases of CJD but have been supplemented in some categories by data obtained from naturally occurring animal TSEs. Information regarding actual infectivity titres in various human tissues other than the brain are extremely limited, but data from experimentally infected animals generally corroborate the grouping shown in the table⁽¹⁾.

The risk of transmission is also influenced by the route of exposure^(24,26). Information regarding human infectivity levels is limited and is extrapolated from experimental research on animals and rare reports of iatrogenic exposure.

Table 3: Tissue Risk for CJD⁽¹⁾	
Level of Infectivity	Tissues, Secretions and Excretions
High Infectivity	Brain, spinal cord, dura mater, pituitary, eye* (including optic nerve and retina)
Low Infectivity	CSF,** kidney, liver, lung, lymph nodes, spleen, placenta ⁽⁸⁸⁾
No Detected Infectivity	Adipose tissue, skin, adrenal gland, heart muscle, intestine, peripheral nerve, prostate, skeletal muscle, testis, thyroid gland, feces, milk, nasal mucus, saliva, serous exudate, sweat, tears, urine, blood, bone marrow, semen

* *The highest levels of infectivity in the eye are associated with the optic nerve and retina and, to a much lower level, the cornea. It is expected that levels of infectivity for other parts of the eye are low or nonexistent. There is no infectivity associated with tears.*

** *Although CSF is classified as low infectivity and is less infectious than high infectivity tissues, it was felt that instruments contaminated by CSF should be handled in the same manner as those contacting high infectivity tissues in high risk and at risk patients⁽¹⁾.*

Dental neurovascular tissue (e.g. dental pulp) has not been assigned a level of infectivity by the WHO consultants, but it may be prudent to treat it as low infectivity tissue in high risk patients. Some experimental studies have demonstrated that TSE-infected animals develop a significant level of infectivity in gingival and dental pulp tissues, and that root canal procedures and gingival abrasions with subsequent exposure to infective brain tissue can transmit TSEs in healthy animals⁽¹⁾.

Risk Management for CJD

A. Infection Prevention and Control Management Based on Risk Assessment for CJD

A risk assessment should be carried out to determine the appropriate infection prevention and control procedures required for CJD. CJD precautions listed in the Recommendations Section (see Part C) are required for very specific invasive clinical situations. CJD precautions should be initiated for a high risk patient when exposure to high or low infectivity tissues is anticipated and for an at risk patient when exposure to high infectivity tissues (including CSF) is anticipated (see Tables 1, 2 and 3).

Normal social and non-invasive contact with a high risk or an at risk patient does not transmit CJD, therefore CJD precautions are not required for these circumstances.

B. Management of Equipment and Environmental Surfaces

Incineration or Decontamination

Guidelines from other countries vary on the recommended method for decontaminating instruments or equipment that have come in contact with high infectivity tissues of patients with CJD. The most stringent recommendations are from the WHO⁽¹⁾, Australia⁽¹³⁾ and the U.K.⁽¹²⁾, where incineration is recommended for instruments exposed to high infectivity tissue. Rutala and Weber have recommended autoclaving rather than incineration⁽⁸⁹⁾.

The CJD agent is extremely hardy and resistant to common decontamination methods^(5,12,13,17,23,87,90), as are all TSEs. Many of the existing decontamination data have come from studies of scrapie and other TSEs in animal populations⁽⁹¹⁻¹⁰⁰⁾. The best defined model is scrapie in mice or hamsters, which has been repeatedly used in studies to establish practical inactivation procedures. Specific parameters addressed have included levels of infectivity after exposure to particular chemicals^(82,101,102), the temperature range for inactivating the agent by steam sterilization, and comparisons between prevacuum and gravity displacement steam sterilizers^(5,87,91,92,103).

Study results have not been consistent^(82,91,102,104). There are conflicting results related to methods of decontamination, including the efficacy of various chemical agents used, the starting titres of infectivity and the amount of host tissue present^(5,82,102,104). Researchers have concluded that even if thermal and chemical methods of decontamination kill most of the infectivity, small refractory subpopulations survive, but the long-term observation needed to establish this may not have been implemented in some decontamination studies^(102,105). The limitations

and conflicting information complicate extrapolation of these findings to the care of humans. Some experimental methods may not be relevant to the human situation:

- ✧ Non-standardized methods with a variety of tissues and varied strains of TSE have been chosen for the experiments (e.g. intact brain, dried macerated brain tissue or brain homogenates)^(5,106).
- ✧ Some of the strains used are more thermostable than others.
- ✧ Varying sample sizes with varying titres of infectivity have been employed.
- ✧ A variety of decontamination studies have used high inoculums and organic matter on instruments not cleaned before the decontamination process.
- ✧ The amount of tissue used frequently exceeded the bioburden normally found on surgical instruments because of the difficulty and extreme expense of conducting low bioburden studies⁽¹⁰⁶⁾.

Incineration or the CJD decontamination process must be followed without exception when instruments are exposed to *high or low infectivity tissues* of a high risk patient or *high infectivity tissues and CSF* of an at risk patient.

Because the instrument is destroyed and cannot be reused, incineration is the safest and most unambiguous method for ensuring that there is no risk of prion transmission by surgical instruments. No decontamination method has been definitely proven to be 100% effective against prions.

To ensure proper functioning and maintenance of an incinerator, the manufacturer's recommendations should be followed. Incineration is a process that converts combustible materials into noncombustible ash, achieving a reduction of 90% by volume or 75% by weight. The product gases are vented into the atmosphere, and the treatment residue may be disposed of in a landfill⁽¹⁰⁷⁾. Waste managers should be consulted for guidance on incineration.

It is *strongly recommended* that reusable instruments used on high infectivity tissues from a high risk or at risk patient be incinerated (see Figures 1 and 2). If incineration is either impossible or impractical (e.g. work surfaces or instruments and equipment that the institution has decided must be reused), the WHO consultation suggests that the combination chemical/sterilization CJD decontamination processes that appear in Table 6 (page 27) should remove most, and possibly all, infectivity under the widest range of conditions. Until studies are published that identify the risk of reusing decontaminated instruments, policy makers are encouraged to adopt the most stringent processing methods possible for CJD⁽¹⁾.

Advance Planning

Advance planning is necessary to limit the need to incinerate or use harsh chemical/steam combinations on delicate, expensive surgical instruments. When surgery on high or low infectivity tissues is planned for a high risk patient, disposable equipment should be used as much as possible. If disposable equipment is unavailable, older equipment near the end of its life span should be used to reduce the cost of instrument destruction and replacement (see Table 4). Expensive heat- and liquid-sensitive equipment may potentially be protected during a surgical procedure by sealing components with temporary disposable covers (i.e. fluid-resistant material). Because of the risk of leakage through the disposable cover, the barrier effectiveness of the cover should be assessed before use and the equipment inspected after surgery.

A detailed inspection of the device should be completed. This inspection may reveal that it can be dismantled, separating heat-sensitive from heat-resistant components. The manufacturer should be contacted for specific information regarding dismantling and decontamination methods relating to the device. Once it has been completely dismantled and thoroughly investigated with respect to decontamination methods, it may be apparent that various parts will tolerate the most stringent chemical CJD decontamination process listed in Table 6, page 27, (i.e. 2.1 or 2.2). If this procedure is completed (i.e. inspection and dismantling), fewer parts may need to be incinerated.

Table 4: Infection Prevention and Control Management Based on Risk Assessment for CJD

Risk Assessment (see Tables 2 & 3)	Prevention Management (risk identified <i>before</i> an invasive procedure)	Containment Management (risk identified <i>during</i> an invasive procedure)	Contingency Management (risk identified <i>after</i> an invasive procedure)
<p>High Risk Patient - involving high or low infectivity tissues or At Risk Patient - involving high infectivity tissues (including CSF)</p>	<ol style="list-style-type: none"> Develop and implement CJD precautions as required for specific departments (e.g. operating suites, laboratory, central processing, etc.) (see Part C). Provide education programs to ensure that personnel are adequately trained. Provide advance notification of admission to appropriate personnel (i.e. infection prevention and control). Provide advance notification to various departments as required (i.e. operating suites, laboratory, central processing, etc.). Consider scheduling surgery at the end of the procedure day. Minimize the number of individuals involved in the procedure. Ensure that only educated and trained individuals are involved. Use disposable supplies and equipment. Keep to a minimum the number of items. If disposable equipment cannot be used, use old instruments that are at the end of their life cycle in set up and procedures. Follow the algorithm for management of devices used on a high risk or at risk patients (see Figures 1, 2 & 3 and Table 6). 	<ol style="list-style-type: none"> Implement CJD precautions immediately (see Part C). Obtain and immediately implement disposable or old instruments that are at the end of their life cycle. Obtain and immediately implement the use of disposable supplies and equipment for invasive procedures. Keep the number of items used to a minimum. Minimize the number of individuals involved in the procedure. Ensure that only educated and trained individuals are involved. Notify appropriate personnel as required (e.g. infection prevention and control, central processing, housekeeping, laboratory, ward and administration). Identify all reusable equipment used on patient prior to the risk being identified. Follow the algorithm for management of devices used on a high risk or at risk patient for CJD (see Figures 1, 2 & 3 and Table 6). 	<ol style="list-style-type: none"> Implement CJD precautions as required (see Part C). Notify appropriate personnel (e.g. administration, infection prevention and control, laboratory, central processing, operating suite) in order to arrange a meeting. Identify reusable equipment exposed to high or low infectivity tissues from a high risk patient or involving high infectivity tissues (including CSF) from an at risk patient (see Tables 2 & 3 and Figures 1, 2 & 3). A detailed inspection of a complex or fragile piece of a reusable device may reveal that the item can be dismantled more thoroughly than anticipated. Various parts of the device may tolerate the CJD decontamination process safely (Table 6). By completing this process, far less of the device may need to be incinerated. The manufacturer can be contacted for assistance. Follow the algorithm for management of devices used on a high risk or at risk patient for CJD (see Figures 1, 2 & 3 and Table 6). A decision whether or not to inform exposed patients will be contingent upon the hospital's policy.
<p>High Risk Patient - involving no detectable infectivity tissues or At Risk Patient - involving low (excluding CSF) or no detectable infectivity tissues</p>	<p>CJD Precautions are not required. Appropriate cleaning and sterilization/disinfection processes are adequate.</p>		

Instrument Quarantine

For reusable instruments used on patients in whom CJD is suspected but not confirmed, instrument quarantine may provide an alternative to incineration or harsh chemical/autoclave decontamination. If a facility can safely quarantine instruments until a diagnosis has been confirmed, quarantine can be used to avoid needless destruction of reusable instruments when suspect cases are later found not to have CJD. Instruments designated to be quarantined must be thoroughly cleaned, kept moist, stored in a puncture-resistant container, dated, and labelled “biohazardous”. A monitoring process must be in place to ensure that the quarantined instruments are not accidentally put into recirculation. If CJD is excluded as a diagnosis, the instruments may be safely returned to circulation after a second thorough cleaning and appropriate sterilization/disinfection process.

Quarantine of instruments after their exposure to high infectivity tissues of an at risk patient may not be realistic because the incubation period may be very lengthy (years).

Non-immersible Equipment

If the reusable instrument cannot be fully immersed in the chemical disinfectant, then it *must be* incinerated.

When a non-immersible, expensive, heat and/or chemical-sensitive device must be used, cover the device by wrapping or sealing it with disposable material (e.g. liquid-resistant materials). After the procedure, incinerate coverings and attachments that could not be wrapped (e.g. drill bits).

Decontamination

Cleaning Phase

Cleaning is an extremely important part of instrument and equipment decontamination and is necessary to permit maximum efficacy of subsequent chemical/heat-steam treatments⁽¹⁰⁷⁾. Recent research suggests that instruments not properly cleaned before chemical/heat-steam treatment may still harbour the CJD agent in the centre of the tissues or fluid remaining on the instruments⁽¹⁰⁸⁾. It is believed that the chemicals or heat-steam may seal the outer layer of the tissue that remains on the instrument, which consequently protects the CJD agent in the core of these particles⁽¹⁰⁹⁾. Effective cleaning can physically remove large numbers of microorganisms⁽¹¹⁰⁾ and thereby remove most of the bioburden prior to the decontamination cycle⁽⁸⁹⁾.

If manual cleaning is used, extreme care should be taken to prevent exposure to contaminated cleaning fluid, especially from power spraying, splashing, and puncture wounds. If an automatic washer is used, on completion of the automatic process the washers should be run through an empty cycle before any further use⁽¹⁾.

The surfaces of all instruments that are going to be reused must be cleaned in advance of chemical or heat decontamination. If the instrument has been disinfected or sterilized and still has visible material on it, then it must be incinerated (dried or baked on tissue will protect the CJD agent from destruction). The instrument must not be rechallenged through the CJD decontamination process.

Chemical Phase

Chemical decontamination includes the use of either of two disinfectants, sodium hypochlorite or sodium hydroxide, which have been found to be effective in decreasing the infectivity of prions^(91-95,99,100,102,111). In one study, an unexplained and unexpected finding with the sodium hydroxide (NaOH) was that 2-hour exposures were less effective against BSE and scrapie agents than exposures of 30 or 60 minutes⁽⁹¹⁾. Both chemicals are irritating to mucous membranes and skin, therefore barrier precautions are required to protect HCWs working with these chemicals. If these chemicals are used as a decontamination method for CJD, they must be disposed of in accordance with provincial regulations (see Appendix III, Material Safety Data Sheets).

HCWs should review the Material Safety Data Sheets (MSDS) before working with these chemicals.

a) Sodium hydroxide (NaOH or soda lye)

Sodium hydroxide is less corrosive than sodium hypochlorite to metals and fabrics, but its effect on any particular instrument cannot be predicted beforehand. It is corrosive to glass and aluminum. Solutions of 1N NaOH should be prepared fresh for each use either from dry NaOH or by dilution from 10N NaOH stock solutions. At room temperature, NaOH is caustic to skin and cloth but is relatively slow acting and can be removed from skin or clothing by thorough rinsing with water. Hot NaOH is aggressively caustic and should never be handled until it has cooled to room temperature.

b) Sodium hypochlorite (NaOCl solution or bleach)

Sodium hypochlorite is extremely corrosive to most metals and fabrics, causing surgical instruments and steel tables to rust and mattress covers to crack. Sodium hypochlorite efficacy depends upon the concentration of available chlorine, and for CJD decontamination NaOCl should be diluted to give a final concentration of 20,000 ppm available chlorine⁽¹²⁾. It is important to remember that concentrated stock solutions must be kept tightly sealed to prevent loss of chlorine due to light contact, and sodium hypochlorite solutions should be prepared fresh for each use. Sodium hypochlorite cannot be used as an instrument bath in a sterilizer.

Heat/Sterilization Phase

Flash and routine steam sterilizing methods are ineffective in destroying the CJD agent and *must never* be considered as a processing method for reusable instruments exposed to the CJD agent.

Gravity displacement and prevacuum (porous load) steam sterilizers have been tested using various temperature and total cycle times^(91-93,95,96,102,111). However, the results are contradictory: some studies report complete inactivation of the prion with gravity displacement and prevacuum sterilizers⁽¹⁰²⁾, and others show incomplete inactivation with prevacuum⁽⁹¹⁾ and gravity displacement steam sterilizers^(92,100). Some experts recommend that gravity displacement should not be used⁽¹²⁾, whereas others indicate only that gravity displacement steam sterilizers are less effective than prevacuum⁽¹³⁾ steam sterilizers for CJD decontamination.

There are no biological indicators to establish successful CJD decontamination. Biological indicators are used to monitor gravity and vacuum assisted steam sterilization cycles to determine that the selected cycle is capable of killing bacterial spores, considered to be the most resistant microorganisms for the process. However, biological indicators containing spores will not indicate that prions are destroyed.

Combination Methods

Studies have demonstrated complete elimination of infectivity using a combination of NaOH followed by steam sterilization with gravity displacement sterilizers^(92,100,111,112).

Ineffective or Partially Effective Methods for CJD

Table 5 lists several chemical disinfectants and sterilization processes according to their effectiveness in reducing infectivity. Activity will vary depending on product formulation, test conditions, process controls and the presence or absence of organic or inorganic soils. It is important to note that after treatment with formalin, the CJD agent becomes resistant to decontamination processes that would otherwise have been effective^(6,93,95,96,102,105,113).

Table 5: Ineffective or Partially Effective Processing Methods for CJD⁽¹⁾	
Chemical Disinfectants	Sterilization
Ineffective* alcohol, ammonia, B-propiolactone, formalin, hydrochloric acid, hydrogen peroxide, peracetic acid, phenolics, formaldehyde, sodium dodecyl sulfate (SDS) (5%)	Ineffective flash or routine steam sterilizing, ethylene oxide, dry heat (< 300° C), ionizing, ultraviolet or microwave radiation, formaldehyde, boiling
Variably or partially effective chlorine dioxide, iodophores, glutaraldehyde, urea (6M), guanidine thiocyanate (4M), sodium dichloroisocyanurate, sodium metaperiodate	Variably or partially effective steam sterilizing at 121° C for > 15 minutes, SDS, boiling in 3% sodium dodecyl

* Some of these chemicals may have very small effects on TSE infectivity, but they are not adequate for disinfection.

Part C. Recommendations

Recommendations for CJD Decontamination Procedures

Table 6 outlines the recommended CJD decontamination processes as adapted from the WHO Infection Control Guidelines for Transmissible Spongiform Encephalopathies, Report of a WHO Consultation, which took place in Geneva, Switzerland during March 23-26, 1999. Because of the difficulties that may be encountered with a sodium hydroxide (NaOH) bath in a steam sterilizer, the concurrent combined method has not been included in these recommendations.

Table 6: CJD Decontamination Processes⁽¹⁾	
<i>These recommendations are based on the best available evidence at this time and are in decreasing order of effectiveness. They must be followed, without exception, when there is exposure to high and low infectivity tissues from a high risk patient and high infectivity tissues and CSF from an at risk patient (see Tables 2 and 3). NOTE: If the instrument or surface cannot be fully immersed or flooded with the chemical disinfectant, then the item must be incinerated.</i>	
1. Incineration: Use for all instruments, effluent materials, and solid waste.	
2. Instrument Decontamination for heat-resistant reusable instruments that an institution is unwilling or unable to incinerate.	
2.1	Cleaning: Removal of adherent particles through mechanical or manual cleaning must be completed prior to any chemical/sterilizer decontamination of instruments. Instruments and other materials to be decontaminated should be kept moist between the time of exposure to infectious materials and subsequent decontamination; then
2.2	Immerse instruments in 1N sodium hydroxide (NaOH) or sodium hypochlorite* solution for 1 hour; remove from chemical solution; rinse instruments well; then immerse instruments in water and place in a sterilizer selecting the liquid cycle and heat to 121° C for 1 hour; or
2.3	Immerse instruments in 1N NaOH or sodium hypochlorite* for 1 hour; remove instruments from chemical solution; thoroughly rinse instruments in water; then transfer to an open pan, place in prevacuum sterilizer, and heat to 134° C for 1 hour or to 121° C in a gravity displacement sterilizer for 1 hour.
3. Hard Surface Decontamination	
3.1	Remove visible soil.
3.2	Flood with 2N NaOH or undiluted sodium hypochlorite; let stand for 1 hour; then mop up and rinse with water; or

*20,000 ppm available chlorine

3.3	If surfaces cannot tolerate NaOH or undiluted sodium hypochlorite, thorough cleaning will remove most infectivity by dilution, and some additional benefit may be derived from the use of one or another of the partially effective methods listed in Table 5.
4. Chemical/Sterilizer Process for Dry Goods	
4.1	Small dry goods that can withstand either NaOH or sodium hypochlorite should first be immersed in one or the other solution (as described in 2.3 above) and then heated in a prevacuum steam sterilizer to 134° C for 1 hour.
4.2	Bulky dry goods or dry goods of any size that cannot withstand exposure to NaOH or sodium hypochlorite should be heated in a prevacuum steam sterilizer to 134° C for 1 hour.

* 20,000 ppm available chlorine

Note: Table 6 recommends that, when possible, two or more different methods of prion inactivation be combined sequentially in any decontamination procedure.

NaOH (or soda lye): Be familiar with and observe safety guidelines for working with NaOH (see MSDS sheet example in Appendix III). 1N NaOH is a solution of 40g NaOH in 1 litre of water. 1N NaOH readily reacts with CO₂ in air to form carbonates that neutralize NaOH and diminish its disinfective properties. 10N NaOH solutions do not absorb CO₂, therefore 1N NaOH working solutions should be prepared fresh for each use either from solid NaOH pellets or by dilution of 10N NaOH stock solution⁽¹⁾. NaOH should be dissolved by adding the pellets carefully. Adding pellets to water may cause spitting, putting the individual preparing the solution at risk of injury. Do not use on aluminum.

Sodium hypochlorite (bleach): Be familiar with and observe safety guidelines for working with sodium hypochlorite (see MSDS sheet example in Appendix III). Household or industrial strength bleach is sold at different concentrations, so that a standard dilution cannot be specified. Its efficacy depends on the concentration of available chlorine and should be 20,000 ppm available chlorine. A common commercial formulation is 5.25% bleach with 52,500 ppm available chlorine⁽⁵⁾. Solutions should be prepared fresh for each use.

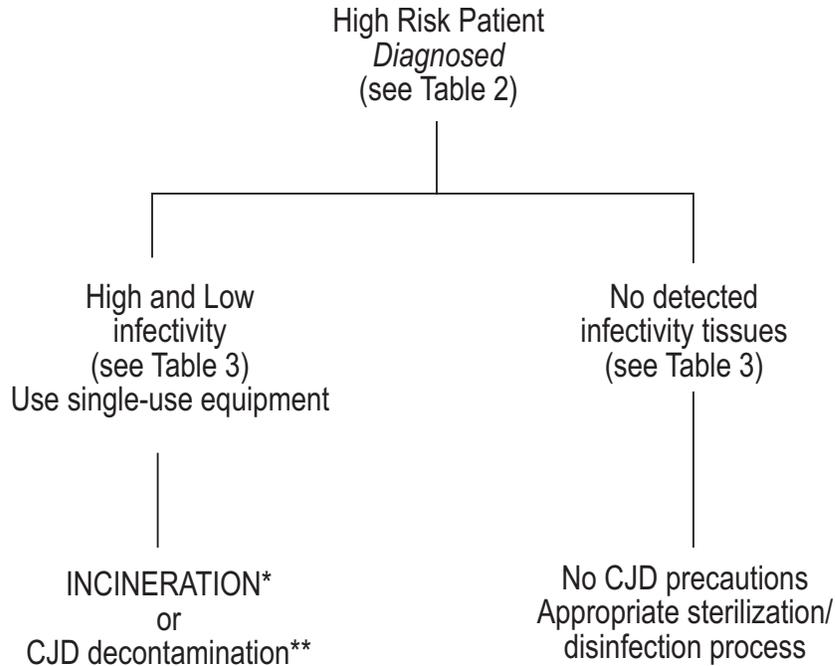
Category B; Grade II
(see Appendix I, page 58,
for Guideline Rating System)

Reusable instruments or instrument components that an institution is unwilling or unable to incinerate must be reprocessed through one of the CJD decontamination processes listed in Table 6. They should then be rechallenged through the appropriate decontamination process used for all reusable instruments (i.e. cleaned again, inspected, function tested, packed and disinfected or sterilized before being made available for use on another patient).

Biological indicators will not indicate that prions are destroyed. Biological indicators are used to monitor the gravity and vacuum assisted steam sterilization cycles to determine that the selected cycle is capable of killing bacterial spores, considered to be the most resistant microorganisms known for the process. However, biological indicators containing bacterial spores will not indicate that prions have been destroyed.

Algorithms for the management of instruments and equipment used on a high risk or an at risk patient are presented in Figures 1, 2 and 3.

Figure 1: Algorithm for the Management of Instruments and Equipment used on a High Risk Patient Diagnosed with CJD (Use single-use instruments and equipment wherever possible.)

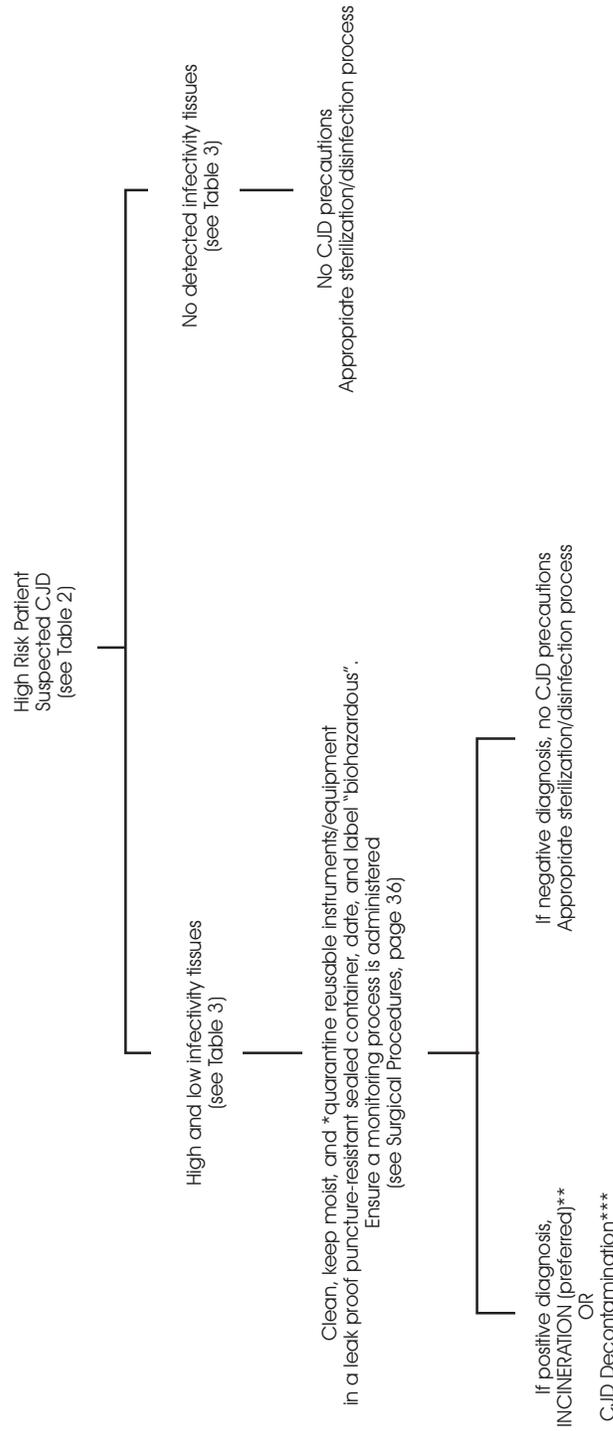


Note: Refer to page 32: Recommendations for the management of a high risk or an at risk patient for CJD in the health care setting for detailed information, in particular D. Medical Procedures and E. Surgical Procedures.

* Complex instruments, e.g. fiberoptic endoscopes, that cannot be adequately decontaminated by the chemical/sterilizer methods as per Table 6 should be incinerated (see Appendix IV).

** Reusable instruments and equipment should be critically assessed for dismantling (see Part B. Risk Management for CJD, Advance Planning, page 21).

Figure 2: Algorithm for the Management of Instruments and Equipment used on a High Risk Patient with Suspected CJD (Use single-use instruments and equipment wherever possible.)



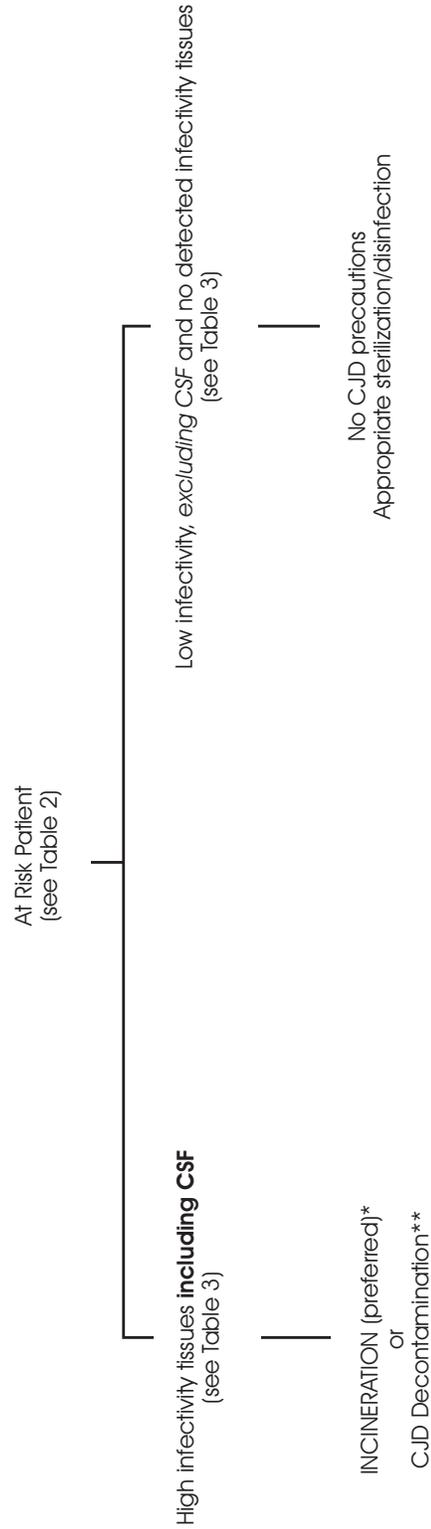
Note: For detailed information refer to page 32: Recommendations for the management of a high risk or an at risk patient for CJD in the health care setting, in particular D. Medical Procedures and E. Surgical Procedures.

* If reusable instruments are not quarantined from a high risk patient in whom CJD is suspected but not confirmed, then they must be treated as if used on a patient with diagnosed CJD (see Figure 1).

** Complex instruments, e.g. fiberoptic endoscopes, that cannot be adequately decontaminated by the chemical/sterilizer methods as per Table 6 should be incinerated (see Appendix IV).

*** Reusable instruments or equipment should be critically assessed for dismantling (see Part B. Risk Management for CJD, Advance Planning, page 21).

Figure 3: Algorithm for the Management of Instruments and Equipment used on an At Risk Patient for CJD (Use single-use instruments and equipment wherever possible for high infectivity tissue and CSF.)



Note: For detailed information refer to page 32: Recommendations for the management of a high risk or an at risk patient for CJD in the health care setting, in particular D. Medical Procedures and E. Surgical Procedures.

* Complex instruments, e.g. fiberoptic endoscopes, that cannot be adequately decontaminated by the chemical/sterilizer methods as per Table 6 should be incinerated (see Appendix IV).

** Reusable instruments or equipment should be critically assessed for dismantling (see Part B. Risk Management for CJD, Management of Equipment and Environmental Surfaces, page 19).

Recommendations for The Management of a High Risk or an At Risk Patient for CJD in the Health Care Setting

Routine Practices (as recommended in *Routine Practices and Additional Precautions for Preventing the Transmission of Infection in Health Care*⁽³⁾) are adequate for providing care to a high risk or an at risk patient in normal social and clinical contacts and for non-invasive clinical investigations^(1,3). CJD precautions apply to GSS and FFI. CJD precautions are recommended for specific clinical situations as outlined below.

A. Administration

- a. At no time should a patient at high risk or at risk for CJD be deferred, denied or in any way discouraged from admission to a health care facility⁽¹⁾

Category B; Grade III

- b. Policy makers are strongly encouraged to develop policies and procedures for infection prevention and control of CJD, including patient risk assessment for CJD prior to any intervention that may involve exposure to high or low infectivity tissues (see Tables 2 and 3).

Category B; Grade III

- c. Policy makers should ensure that policies and procedures are developed with input from infection prevention and control as well as occupational health personnel.

Category B; Grade III

- d. Policy makers are encouraged to adopt the most stringent CJD decontamination processes possible, as listed in Table 6, until further information is available to clarify the risk of reusing decontaminated instruments or equipment previously exposed to the CJD agent⁽¹⁾.

Category B; Grade III

- e. Administration should define who has the authority to determine whether a device undergoes incineration, recognizing that the infection prevention and control personnel may have the most expertise to make such a recommendation.

Category B; Grade III

- f. Each health care facility should identify how it will incinerate items that will require incineration.

Category B; Grade III

- g. Each health care facility should identify a laboratory to which it can refer specimens for the diagnosis of CJD.

Category B; Grade III

- h. Each health care facility is strongly encouraged to devise a system for tracking instruments and equipment used in neurosurgical, spinal and ophthalmologic procedures involving high infectivity tissues and CSF.

Category B; Grade III

B. Notification

- a. It is the responsibility of the attending physician or surgeon to provide advance notification to the health care facility if a high risk or an at risk patient is to be admitted (see Tables 2 and 4).

Advance notification should include

1. administration and Infection Prevention and Control Program personnel on admission;
2. operating suites, pathology, laboratory, infection prevention and control personnel, and Central Processing, before any medical or surgical procedures are carried out that may involve exposure to high or low infectivity tissues (see Table 3);
3. all patient care areas such as critical care units, step-down units and general wards, in order to implement CJD precautions when contact with high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF of an at risk patient is possible (e.g. use of disposable set ups for central nervous system monitoring and drainage systems);
4. the morgue prior to an autopsy.

Category B; Grade III

- b. The health care facility is responsible for notifying funeral homes or other organizations that may be handling high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient (e.g. in mortuary procedures, laboratory investigations).

Category B; Grade III

- c. Patients at high risk or at risk for transmission of CJD or their responsible care giver should be advised to notify their doctors, dentists and other health care workers of their status so that CJD precautions can be implemented as required.

Category B; Grade III

C. General Patient Care

- a. Routine Practices⁽³⁾ apply to all care procedures except for invasive procedures involving high or low infectivity tissue of a high risk patient or high infectivity tissues and CSF of an at risk patient (see Tables 1 and 2).

Category B; Grade III

- b. A single room is not required for infection prevention and control purposes^(1,12).

Category B; Grade III

- c. No CJD precautions are required for feeding utensils, feeding tubes, suction tubes, razors, or items used in the care of skin or bed sores. Contamination from tissues with no detected infectivity (see Table 3) poses no greater hazard than for any other patient.

Category B; Grade III

- d. Routine Practices⁽³⁾ apply to most situations involving linen. Linen (bed sheets, operating room linens, personal protective equipment) that are exposed to high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF of an at risk patient (see Tables 2 and 3), should be sealed in a leak proof container, labelled “biohazardous”, and incinerated, or submitted to CJD decontamination processes as identified in Table 6.

Category B; Grade III

D. Medical Procedures

High risk or at risk patients may need to undergo diagnostic procedures unrelated to CJD. In general, these procedures may be conducted without any special precautions, as long as procedures only involve tissues containing no detected infectivity⁽¹⁾ (see Table 3). The following recommendations should be followed for the specific medical procedures outlined below.

- a. During invasive medical interventions that may involve the collection and handling of high or low infectivity specimens of a high risk patient or high infectivity specimens and CSF of an at risk patient (e.g. during lumbar puncture or when taking CSF samples from an external ventricular drain), CJD decontamination of instruments as per Table 6 must be followed^(12,13) as well as CJD precautions for collecting and handling specimens (see J. Laboratory, page 43).

Category B; Grade III

- b. Endoscopy equipment (e.g. bronchoscope) used on high or low infectivity tissues of a high risk patient in whom CJD has been diagnosed or on high infectivity tissues and CSF of an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated^(1,13,114) (see Figures 1 and 3), as it is not possible to guarantee adequate decontamination (see Appendix IV). Accessories that do not come into contact with the patient

(e.g. cameras) should be protected from surface contamination by wrapping or bagging as much of the instrument as possible with disposable liquid-resistant materials that can later be removed. Wrapping/bagging material should be incinerated (see I. Waste Disposal, page 42).

Category B; Grade III

- c. Endoscopy equipment used on high or low infectivity tissues of a high risk patient in whom CJD is suspected but not confirmed should be thoroughly cleaned, kept moist and quarantined without undergoing CJD decontamination or routine disinfection/sterilization. The instrument should be stored in a dated, leak proof, puncture-resistant, sealed container and labelled “biohazardous.” The container should then be stored in a secure area and a monitoring process should be in place to ensure that the instrument is not recirculated into the system until the diagnosis has been confirmed by neuropathological examination. If the diagnosis is positive for CJD, the instrument should be incinerated (see Figure 2 and Appendix IV)⁽¹⁾. If the diagnosis is not CJD, the instrument does not require CJD precautions, and routine cleaning and sterilization/disinfection may be initiated (see Figure 2).

Category B; Grade III

- d. Endoscopy equipment used on tissues with no detected infectivity from a high risk or an at risk patient (e.g. during gastric/intestinal endoscopy) or on the low infectivity tissues (excluding CSF) of an at risk patient does not require CJD precautions (see Figures 1, 2 and 3).

Category B; Grade III

- e. All intracerebral EEG electrodes used on a high risk or an at risk patient must be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated⁽⁷²⁾.

Category A; Grade II

- f. Surface skin electrodes for electroencephalography (EEG) do not require CJD precautions, except when the CNS tissue or fluid of a high risk or an at risk patient is exposed (e.g. during neurosurgical procedures, trauma). In such cases, the EEG electrodes should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated.

Category B; Grade III

- g. No CJD precautions are required for electromyography (EMG) needles.

Category B; Grade III

- h. Non contact air or puff tonometers that totally avoid contact with the cornea should be used whenever possible⁽¹³⁾. However, if these are not available, disposable, plastic tonometer covers must be used. The covers should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated after use⁽¹³⁾. Tonometers, suction cups used in laser procedures, and other instruments that come into direct contact with the cornea of a high risk or

an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated^(13,115).

Category B; Grade III

- i. No CJD precautions are required for hemodialysis machines.

Category B; Grade III

E. Surgical Procedures

1. **High risk patient** (involving high or low infectivity tissues) **OR at risk patient** (involving high infectivity tissues, including CSF – see Table 4 and Figures 1, 2 and 3)

Surgical procedures involving high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF of an at risk patient require CJD precautions. These procedures include, but are not limited to, neurosurgical and spinal procedures that involve opening the dura mater (including dura mater graft procedures and use of intracerebral EEG electrodes), ophthalmic procedures, and situations of trauma in which CNS tissue or fluid is exposed. The following recommendations should be followed.

- a. The number of HCWs involved in the surgery should be limited to the minimum required^(1,12,13). Staff should be educated and trained about CJD precautions and the occupational risks and hazards related to CJD^(1,12,13).

Category B; Grade III

- b. HCWs should wear appropriate single-use personal protective equipment^(12,13,106), such as gloves, liquid-repellent gowns, fluid-resistant aprons, head and foot coverings, face shields, and masks^(1,116). Protective eye wear (e.g. safety glasses, visor, or goggles) or other protective devices must be worn when splashes or particles are possible to the eyes and the face^(1,11-13,106). Personal protective equipment (single use or non-disposable) exposed to high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated^(1,12,13,106) or decontaminated as per Table 6.

Category B; Grade III

- c. Single use items should be used whenever possible^(1,12,13,106)

Category B; Grade III

- d. Supplies and equipment should be limited to only what is necessary for the procedure⁽¹⁰⁶⁾.

Category B; Grade III

- e. Temporary covers, shields, or guards made of disposable, liquid-resistant materials that can be removed and incinerated should be used wherever surfaces such as surgical tables, counter tops, instruments (e.g. drills) or equipment might be exposed to high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient^(1,12,106).

Category B; Grade III

- f. One-way flow of instruments should be maintained⁽¹⁾. Sharp instruments should be passed in trays, not from hand to hand⁽¹¹⁷⁾.

Category B; Grade III

- g. Instruments exposed to high or low infectivity tissues should be separated from instruments exposed to no detected infectivity tissues⁽¹⁾

Category B; Grade III

- h. It is strongly recommended that instruments that have been exposed to high infectivity tissues and CSF from a high risk patient with a diagnosis of CJD^(1,12) or from an at risk patient be incinerated (see Figures 1 and 3). Incineration is also preferred for instruments that have been exposed to low infectivity tissues from a patient with a CJD diagnosis (see Figure 1). Instruments and equipment parts that are to be incinerated need to be sealed in a leak proof, puncture-resistant container and labelled “biohazardous”⁽¹³⁾. If incineration is not possible, instruments should be decontaminated as per Table 6.

Category B; Grade III

- i. Reusable instruments and equipment exposed to high or low infectivity tissues from a high risk patient in whom CJD is suspected but not confirmed should be thoroughly cleaned, kept moist and quarantined, but not subjected to CJD decontamination or routine disinfection/sterilization. The instruments are to be stored in a dated, leak proof, puncture-resistant, sealed container and labelled “biohazardous.” The container should be stored in a secure area, and a monitoring process should be in place to ensure that the instruments are not recirculated into the system until the diagnosis has been confirmed by neuropathological examination. If the diagnosis is positive for CJD, the instruments should be incinerated^(1,12,13,86,118) or decontaminated as per Table 6. If the diagnosis is not CJD, the instruments do not require CJD precautions, and routine cleaning and sterilization/disinfection may be initiated (see Figure 2).

Category B; Grade III

- j. Stereotactic CNS procedures are not recommended for high risk and at risk patients. If stereotactic biopsy is done, instruments exposed to high infectivity tissues and CSF should be managed as in recommendations h. and i. (above). Under normal circumstances, the screws and frame components are not exposed in this way. However, screws that penetrate the skull should be managed as instruments exposed to high infectivity tissues and CSF, as in recommendations h. and i. (above); the same procedures

should be followed if the frame components become contaminated when penetrating screws are removed.

Category B; Grade III

- k. High or low infectivity specimens of a high risk patient, or high infectivity specimens and CSF of an at risk patient that are sent to the laboratory should be sealed in a leak proof, puncture-resistant container and clearly labelled as high risk for CJD^(12,13).

Category B; Grade III

- l. To facilitate cleaning and decontamination of the operating suite, surgical procedures performed on high risk or at risk patients may be carried out at the end of the day's operating list^(1,12,13) (see Table 6 for surface cleaning recommendations).

Category B; Grade III

- m. All solid waste exposed to high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF of an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled "biohazardous," and incinerated (see also I. Waste Disposal, page 42).

Category B; Grade III

- n. Clean and decontaminate all surfaces that are exposed to high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF of an at risk patient, according to the CJD Decontamination Processes, Table 6 (see also H. Environmental Surfaces, page 42).

Category B; Grade III

2. **High risk patient** (involving no detected infectivity tissues) **OR at risk patient** (involving low infectivity, *excluding CSF*, or no detected infectivity tissues, see Figures 1, 2 and 3)

CJD precautions are not necessary, and routine cleaning and sterilization/disinfection processes are adequate.

Category B; Grade III

F. Pregnancy/Childbirth

CJD is not known to be transmitted from mother to child during pregnancy and childbirth. The following recommendations should be followed for labour and delivery. Recommendations for other invasive medical or surgical procedures are found in previous sections of this guideline.

- a. In general, childbirth for women at high risk or at risk for CJD should be managed using Routine Practices⁽³⁾. Because the placenta is considered low infectivity tissue (see Table 2), CJD precautions should be taken for high risk women to reduce the risk of exposure to placenta and other products of conception.

Category B; Grade III

- b. Staff should be educated and trained about CJD precautions, and the occupational risks and hazards related to CJD.

Category B; Grade III

- c. HCWs should wear appropriate single-use personal protective equipment such as gloves, liquid-repellent gowns, fluid-resistant aprons, head and foot covering, face shields, and masks. Protective eye wear (e.g. safety glasses, visor, or goggles) or other protective devices must be worn when it is necessary to protect the eyes and face from splashes and particles.

Category B; Grade III

- d. The placenta and products of conception of women at high risk for CJD must be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and disposed of by incineration. The placenta and products of conception *must not* be used for any other purpose.

Category B; Grade III

- e. If the placenta or products of conception are to be sent to the laboratory for investigation, they should be sealed in a leak proof, puncture-resistant container and clearly labelled as high risk for CJD.

Category B; Grade III

- f. Instruments and equipment exposed to the placenta and products of conception of women at high risk for CJD should be decontaminated by one of the CJD decontamination processes as outlined in Table 6 (see also Figures 1 and 2).

Category B; Grade III

- g. Linen, solid waste, personal protective equipment (single use or non-disposable) exposed to the placenta and products of conception of women at high risk for CJD should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous,” and incinerated or submitted to CJD decontamination as per Table 6.

Category B; Grade III

- h. Environmental surfaces exposed to the placenta or products of conception of women at high risk for CJD should be cleaned and decontaminated by one of the CJD decontamination processes as per Table 6 (see also H. Environmental Surfaces, page 42).

Category B; Grade III

- i. No CJD precautions are required for the newborn.

Category B; Grade III

G. CJD Decontamination Process for Instruments and Equipment

- a. Only experienced people educated and trained in the process of cleaning and disinfection/sterilization should be performing these functions⁽⁴⁾. Staff should be educated and trained about CJD precautions, and the occupational risks and hazards related to CJD^(1,4,12,13,84).

Category B; Grade II

- b. HCW should be familiar with and observe safety guidelines when working with hazardous chemicals such as sodium hydroxide and sodium hypochlorite (see Appendix III for MSDS examples).

Category B; Grade II

- c. HCWs cleaning and handling instruments in the CJD decontamination process (e.g. Central Processing personnel) should wear appropriate single-use personal protective equipment^(12,13,106), such as gloves, liquid-resistant garments with sleeves, either a full face shield or a fluid-impervious face mask and protective eye wear (e.g. safety glasses, visor, or goggles)^(2,4). HCWs should cover skin wounds, eczematous lesions, etc. with waterproof dressings before putting on gloves⁽¹³⁾. Personal protective equipment (single use or non-disposable) exposed to fluids from instruments or equipment potentially contaminated with CJD should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated^(1,12,13,58). Alternatively, non-disposable personal protective equipment may be submitted to CJD decontamination as per Table 6.

Category B; Grade II

- d. Cover work surfaces with disposable liquid-resistant materials, which can then be removed, sealed in a leak proof container, labelled “biohazardous”, and incinerated; otherwise clean and decontaminate underlying surfaces thoroughly as outlined in Table 6 (see also H. Environmental Surfaces, page 42).

Category B; Grade III

- e. Reusable instruments should be kept moist prior to initiating the CJD decontamination process or while quarantined⁽¹⁾.

Category A; Grade II

- f. Clean and decontaminate reusable instruments that have been exposed to high or low infectivity tissues separately from those reusable instruments that have been exposed to no detected infectivity tissues⁽¹⁾.

Category B; Grade II

- g. Reusable instruments should be cleaned as soon as possible after use to minimize drying of tissues, blood, and body fluids on the item⁽¹⁾.

Category B; Grade III

- h. Reusable instruments should be manually cleaned using an enzymatic cleaner prior to CJD decontamination.

Category B; Grade III

- i. Reusable instruments to be cleaned in an automatic mechanical processor must be manually cleaned before they are put in the processor⁽⁴⁾. After the instruments have been through the processor, the mechanical washer/disinfector should be run through a complete empty cycle before any further use⁽¹⁾ (see also I. Waste Disposal, page 42).

Category B; Grade III

- j. For reusable instruments and equipment exposed to high or low infectivity tissues from a high risk patient in whom CJD is suspected but not confirmed, instrument quarantine may provide an alternative to incineration or harsh chemical/heat decontamination. Instruments to be quarantined must be thoroughly cleaned, kept moist, stored in a leak proof, puncture-resistant, sealed container, dated, and labelled “biohazardous”. They should not be subjected to routine sterilization/disinfection. A monitoring process must be in place to ensure that the quarantined instruments are not accidentally put into recirculation. If CJD is excluded as a diagnosis, the instruments may be safely returned to circulation after a second thorough cleaning and appropriate sterilization/disinfection process. Quarantine of instruments after being exposed to high infectivity tissues and CSF of an at risk patient may not be realistic because the incubation period of CJD may be very lengthy (years).

Category B; Grade III

- k. If the CJD decontamination processes do not thoroughly clean the reusable instrument (e.g. visual inspection reveals adherent material still on it), the instrument must be incinerated. The reusable instrument *must not* be re-challenged through the CJD decontamination processes a second time.

Category B; Grade III

- l. Upon completion of the cleaning procedure, all solid wastes, including disposable cleaning materials, should be collected and decontaminated. Incineration is highly recommended (see also I. Waste Disposal, page 42). The cleaning station should then be decontaminated as per Table 6 (see also H. Environmental Surfaces, page 42).

Category B; Grade III

H. Environmental Surfaces

- a. Surfaces (e.g. floors, counter-tops) exposed to high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient should be cleaned and decontaminated by a CJD decontamination process outlined in Table 6^(1,13). When possible, efforts should be made to prevent contamination to surfaces (i.e. through the use of temporary covers, shields, or guards made of disposable, liquid-resistant materials that can then be removed, sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated).

Category B; Grade III

- b. Spills of fluids containing high or low infectivity tissue from high risk patients or high infectivity tissue and CSF from at risk patients should be flooded with full strength sodium hypochlorite for 10 minutes before the absorbent material is applied⁽¹⁾. After the absorbent material has been used to remove the fluid it should then be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated. The surface should then be disinfected using a CJD decontamination process (see Table 6).

Category B; Grade III

I. Waste Disposal

- a. All solid waste exposed to high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated.

Category B; Grade III

- b. Liquids used for cleaning can be flushed down the drain.

Category B; Grade III

- c. Employees should use personal protective equipment (e.g. full face shields) and engineering controls (e.g. splash guards) to prevent exposure from splashing and aerosols during the emptying of waste containers.

Category B; Grade III

J. Laboratory

1. Specimen collecting and handling

CJD precautions are warranted when collecting and handling high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient (see Tables 2 and 3). The following recommendations are to be applied.

- a. Staff should be educated and trained about CJD precautions, and the occupational risks and hazards associated with collecting and handling high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient^(1,12,119).

Category B; Grade II

- b. HCWs should wear appropriate single-use personal protective equipment, such as gloves, gowns, and masks, when collecting and handling high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient⁽¹⁾.

Category B; Grade II

- c. Protective eye wear (e.g. safety glasses, visor, or goggles) or other protective devices must be worn when it is necessary to protect the eyes and face from splashes and particles⁽¹⁾. Personal protective equipment (single use or non-disposable) exposed to high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled “bio-hazardous”, and incinerated^(1,12,13,106) or submitted to CJD decontamination processes as per Table 6.

Category B; Grade III

- d. High or low infectivity specimens from a high risk patient or high infectivity specimens and CSF from an at risk patient should be sent to the laboratory in a sealed, leak proof, puncture-resistant container that is clearly labelled as high risk for CJD^(12,13).

Category B; Grade III

- e. Single-use instruments should be used when performing lumbar punctures on a high risk or an at risk patient to obtain CSF samples or when managing drainage systems such as those used with external lumbar and ventricular drains, and intracranial monitoring systems^(12,13).

Category B; Grade III

- f. A sealed container and closed centrifuge bucket must be used when mixing or when centrifugation is to be carried out with high or low infectivity tissue of a high risk patient or high infectivity tissue and CSF of an at risk patient⁽¹³⁾. The use of a biological safety cabinet is recommended for opening the container⁽¹³⁾. Specimens should be sealed in a leak proof,

puncture-resistant container, labelled “biohazardous”, and disposed of by incineration^(12,13).

Category B; Grade II

- g. All single-use laboratory instruments or equipment used on high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated.

Category B; Grade III

- h. All non-disposable laboratory instruments or equipment used on high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient (e.g. hemocytometers) may be cleaned and decontaminated as per Table 6. No special measures are required for cleaning or decontaminating auto-analysers.

Category B; Grade III

- i. Laboratory work surfaces that have been exposed to high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient should be thoroughly cleaned and decontaminated as per Table 6 (see also H. Environmental Surfaces, page 42).

Category B; Grade III

- j. All laboratory waste exposed to high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF of an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated (see I. Waste Disposal, page 42).

Category B; Grade III

- k. CJD precautions are not necessary for collecting, handling, and processing of blood, urine, and fecal specimens^(11,12,119).

Category B; Grade III

2. Pathology (See K for autopsy recommendations, page 46)

It is essential to ensure that specimens are handled safely and that exposure of the work site and cross-contamination of other specimens are avoided. CJD precautions are warranted when working with high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient (see Tables 2 and 3), because these tissues present a risk of exposure to the CJD agent⁽¹²⁾. The following recommendations are to be applied

- a. Staff should be educated and trained about CJD precautions and the occupational risks and hazards when working with high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient⁽¹⁾.

Category B; Grade II

- b. When handling high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient, particular care should be taken to avoid accidental inoculation or injury. Laboratory staff should wear appropriate single-use personal protective equipment, such as liquid-repellent gowns over plastic apron, gloves (cut-resistant gloves are preferred for brain cutting), masks, and protective eye wear (e.g. safety glasses, visor, or goggles)^(1,12,13). Personal protective equipment (single use or non-disposable) exposed to high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated^(1,12,13,106) or decontaminated as per Table 6.

Category B; Grade II

- c. A separate area with a biological safety cabinet should be available for cut-up or blocking of tissue samples (high and low infectivity issues of a high risk patient and high infectivity tissues and CSF of an at risk patient)^(13,120).

Category B; Grade II

- d. To help reduce the risk of handling high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient, these tissues should be fixed in formalin, immersed in formic acid (> 95%) for one hour^(12,13,121,122) and then transferred to fresh formalin for at least 48 hours before further processing⁽¹⁾. Sections should not be frozen.

Category A; Grade II

- e. Slides made from sections that have been treated with formic acid can be considered non-infectious. Slides made from sections that have not been treated with formic acid may be handled as per laboratory safety practices⁽¹¹⁹⁾, when the cover slip is sealed to the slide and chemically disinfected to ensure external sterility. These slides should be labelled “biohazardous”. If any of the slides are damaged they should be incinerated⁽¹⁾.

Category B; Grade II

- f. Single-use laboratory instruments and equipment should be used wherever possible. Microtomy should be performed using disposable blades. All items exposed to high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF from an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled “biohazardous”, and incinerated.

Category B; Grade III

- g. Surfaces of non-disposable laboratory instruments and equipment should be subject to stringent CJD decontamination as listed in Table 6, in order to prevent cross-contamination of tissue samples⁽¹²⁾.

Category B; Grade III

- h. Laboratory work surfaces that have been exposed to high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient should be thoroughly cleaned and then put through one of the CJD decontamination processes listed in Table 6 (see also H. Environmental Surfaces, page 42).

Category B; Grade III

- i. All the processing fluids that are collected in special waste containers, wax shavings and specimens of high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient should be sealed in a leak proof, puncture-resistant container, labelled “bio-hazardous,” and incinerated⁽¹²⁾ (see also I. Waste Disposal, page 42).

Category B; Grade III

K. Autopsy

Postmortem examination remains an essential element in confirming the clinical diagnosis and the cause of death as CJD⁽¹⁾. The following recommendations are to be applied for high risk or at risk CJD cadavers.

1. General considerations

- a. Some experts recommend that all autopsies on high risk or at risk CJD cadavers should be performed in a high risk autopsy suite at a designated facility⁽¹³⁾, whereas others suggest that it is not essential to have a separate high risk suite⁽¹²³⁾. It may be advisable to have designated centres with skilled professionals in each health care region.

Category B; Grade III

- b. Anatomy departments should not accept, for teaching or research purposes, any body or organs from a high risk or an at risk patient for CJD, unless they have specific training or research programs for CJD, including access to specialized equipment, procedures, appropriate containment facilities and training in the management of high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient.

Category B; Grade III

2. Transport of a high risk or at risk CJD cadaver

- a. A policy should be established that outlines the procedure needed to arrange transportation of the body to the high risk autopsy site. All bodies should be transported inside an impervious body bag and placed in a designated morgue space^(12,120,121). If the skull is open or there is CSF leakage and sutures do not completely control this leaking, a sealed body bag lined with materials to absorb the fluids should be used⁽¹⁾.

Category B; Grade III

- b. The body must be identified by a bracelet or toe tag, on which the CJD risk is noted, and be accompanied by appropriate documentation.

Category B; Grade III

3. Performance of an autopsy on a high risk or at risk CJD cadaver

- a. A protocol for performance of autopsies and safe removal of nervous tissue should be established, supported by resources and training, and communicated to all involved personnel.

Category B; Grade III

- b. The autopsy should be performed by experienced autopsy suite staff^(12,13) who are educated and trained about CJD precautions, and the occupational risks and hazards related to CJD^(12,13).

Category B; Grade III

- c. The autopsy should be restricted to working hours when two autopsy assistants or technicians (or medical technologists) are available to assist with the procedure⁽¹²⁾. Whenever possible, the number participating in the autopsy should be limited to the pathologist and two autopsy staff. One staff member should assist with the prosection while the other performs clean tasks (e.g. handling supplies).

Category B; Grade III

- d. Appropriate single-use personal protective equipment, such as double gloves, gowns, waterproof aprons, masks, and protective eye wear^(1,85), is required for all autopsy staff when performing an autopsy on a high risk or an at risk patient for CJD⁽¹⁾. Those who participate in prosection should wear chain mail⁽⁸⁶⁾ or cut-resistant gloves between two disposable gloves⁽¹²³⁾ on their non-cutting hand.

Category A; Grade II

- e. A set of reusable instruments should be dedicated for use on high or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient^(1,12,121). The instruments should be cleaned and submitted to CJD decontamination (see Table 6), and be separated and clearly labelled “CJD instruments”. They may be reused on subsequent CJD cadavers.

Category A; Category B; Grade III

- f. The autopsy table should be covered with impermeable absorbent pads. The skull can be opened by using the plastic bag technique to ensure that bone dust and CSF are contained^(1,120,123).

Category B; Grade III

- g. Manual saws are recommended in order to avoid the creation of tissue particles and aerosols, and for ease of decontamination after use. Electric saws, if used, should be operated inside an aerosol-containing bag unless ventilated helmets with an appropriate filter are worn⁽¹⁾.

Category B; Grade III

- h. The brain should be placed in a pre-weighed container and weighed in the container to avoid contamination of the scale.

Category B; Grade III

- i. High or low infectivity specimens of a high risk patient or high infectivity specimens and CSF of an at risk patient that are sent to the laboratory should be sealed in a leak proof, puncture-resistant container and clearly labelled as high risk for CJD.

Category B; Grade III

- j. General autopsy dissection, if required, should be performed in block. Water should be turned off to minimize production of contaminated fluids. The organ block should be placed on absorbent pads on the autopsy table and dissection kept to a minimum. Organs should not be weighed and should be returned to the body cavity.

Category B; Grade III

- k. On completion of the autopsy, the body should be closed, washed with water, placed in the impervious body bag, sealed and returned to the morgue.

Category B; Grade III

- l. All the disposable material (e.g. personal protective equipment, absorbent pads) and other wastes should be sealed in leak proof containers, labelled “biohazardous”, and incinerated⁽¹²⁾ (see also I. Waste Disposal, page 42).

Category B; Grade III

- m. Brain specimens and other residual tissues can be discarded by draining the formalin into fluid waste containers for incineration. Brain specimens should remain in their containers and be placed in leak proof bags for incineration. Fluid waste can be absorbed in sawdust and incinerated as solid waste.

Category B; Grade III

- n. Working surfaces (e.g. the autopsy table) should be thoroughly cleaned and decontaminated by one of the CJD decontamination processes as per Table 6 (see also H. Environmental Surfaces, page 42).

Category B; Grade III

Recommendations for The Management of a High Risk or an At Risk Patient for CJD in Community Health Care Settings

(e.g. Home Care Settings, Alzheimer Units, Physicians' Offices)

Routine Practices⁽³⁾ are adequate for the management of a high risk or an at risk patient for CJD in community health care settings⁽¹²⁴⁾, except during specific medical procedures (see D. Medical Procedures, page 34).

Category A; Grade III

Recommendations for Dentistry for The Management of a High Risk or an At Risk Patient for CJD

Epidemiologic investigations have not revealed any evidence that dental procedures lead to increased risk of iatrogenic transmission of CJD among humans. However, some experimental studies have demonstrated that TSE-infected animals develop a significant level of infectivity in gingival and dental pulp tissues, and that root canal procedures and gingival abrasions with subsequent exposure to infective brain tissue can transmit TSEs in healthy animals⁽¹⁾. Therefore, patients at high risk for CJD or their responsible care giver should notify the dentist before all dental procedures, in order for CJD precautions to be implemented as required.

The following are general recommendations to assist dental workers when performing dental procedures on high risk patients for CJD.

- a. Staff should be educated and trained about CJD precautions, and the occupational risks and hazards related to CJD⁽¹³⁾.

Category B; Grade III

- b. Dentists should take an appropriate medical history of all their patients⁽¹³⁾, including a risk assessment for CJD, before all dental procedures (see Table 2).

Category B; Grade III

- c. General infection control practices are sufficient when treating high risk patients during procedures not involving neurovascular tissue⁽¹⁾.

Category B; Grade III

- d. Dental procedures in which neurovascular tissues (e.g. dental pulp) on a high risk patient may be exposed should be carried out at appropriate referral centres by personnel familiar with CJD precautions^(12,13).

Category B; Grade III

- e. Dental staff should wear appropriate single-use personal protective equipment such as gloves, gowns, mask, and protective eye wear or chin-length plastic face shields during procedures in which splashing or spattering is anticipated, likely during all dental procedures^(13,85). Personal protective equipment (single use or non-disposable) exposed to neurovascular tissues (e.g. dental pulp) from a high risk patient should be sealed in a leak proof container, labelled “biohazardous”, and incinerated or decontaminated as per Table 6.

Category B; Grade III

- f. Only single-use needles, syringes and anesthetic cartridges should be used^(1,13,85).

Category B; Grade III

- g. In a high risk patient, neurovascular tissues should be treated as low infectivity tissue. Such neurovascular tissues should be sealed in a leak proof container, labelled “biohazardous”, and incinerated.

Category B; Grade III

- h. Instruments such as hand pieces, dental broaches, burrs and other items exposed to neurovascular tissue from a high risk patient should be disposed of in a sealed, leak proof container, labelled “biohazardous”, and incinerated⁽¹³⁾. This recommendation does not extend to the drill itself, if it has been covered with disposable, liquid-resistant materials during splattering procedures.

Category B; Grade III

- i. Single-use instruments and equipment should be used whenever possible on a high risk patient⁽¹³⁾.

Category B; Grade III

- j. Dentists who routinely perform dental procedures on patients with dementia should retain equipment dedicated for use on the subpopulation of high risk patients.

Category B; Grade III

- k. Dental procedures involving neurovascular tissue on high risk patients may be carried out at the end of the day to permit more extensive cleaning and decontamination⁽¹⁾.

Category B; Grade III

- l. All waste exposed to neurovascular tissue from a high risk patient should be sealed in a leak proof container, labelled “biohazardous”, and incinerated.

Category B; Grade III

- m. Working surfaces exposed to neurovascular tissue from a high risk patient should be cleaned and decontaminated by the CJD decontamination process.

Category B; Grade III

- n. For dental instruments and items in contact with all tissues from an at risk patient, CJD precautions are not required, and routine cleaning and disinfection/sterilization methods are adequate⁽¹⁾.

Category B; Grade III

Recommendations for Funeral Service Workers in The Management of Human Remains from a High Risk Patient for CJD

Concern about possible unknown CJD cases does not warrant an enhanced level of precautions for undertakers handling intact bodies over the precautions used generally for all work⁽¹²⁾. The following are general recommendations to assist funeral service workers when they perform mortuary procedures on the human remains of a high risk patient (see Table 2), to ensure the safety of the personnel and avoid contamination of the workplace.

- a. Embalmers and funeral industry workers should be educated and trained about CJD precautions, and occupational risks and hazards related to CJD.

Category B; Grade III

- b. The unembalmed body should be transported to the preparation room in a sealable, impermeable plastic pouch.

Category B; Grade III

- c. Embalming of an intact, unautopsied body of a high risk patient can be managed safely when CJD precautions are followed. Canadian funeral homes that are unable to maintain CJD precautions should not perform the embalming.

Category B; Grade III

- d. Occupational exposure to the human remains of a high risk patient should be minimized, especially when the body has been autopsied or has sustained traumatic injury⁽¹²⁾ affecting high or low infectivity tissues.

Category B; Grade III

- e. Ordinary contact or handling of human remains of a high risk patient, on which no autopsy has taken place, does not pose a risk of exposure to the CJD agent. When dealing with intact human remains, cosmetic work may be undertaken without using special precautions^(1,12).

Category B; Grade III

- f. Relatives of the deceased should not be discouraged from superficial contact with the body of a high risk patient, such as touching or kissing the face⁽¹⁾.

Category B; Grade III

- g. A steel casket or case is not necessary.

Category B; Grade III

- h. There are no extra precautions required for either burial or cremation⁽¹²⁾. Interment in a closed coffin does not present any significant risk of environmental contamination, and there is no risk of residual infectivity after cremation⁽¹⁾.

Category B; Grade III

- i. If a body of a high risk patient needs to be exhumed or disinterred, the normal standard practice for this procedure should be followed. The body will have the same infectivity as at the time of burial, and the CJD precautions implemented for performing an autopsy should be followed.

Category B; Grade III

- j. Cosmetic work on an autopsied or traumatized body of a high risk patient is not encouraged. If done, care should be taken to limit contamination of the workplace by any leaking bodily fluids from the cranium.

Category B; Grade III

- k. Embalming an autopsied or traumatized body of a high risk patient is not encouraged. If done, the following additional CJD precautions are to be strictly maintained.

- ✧ Embalming should be performed by licensed, experienced embalmers, who are educated and trained about CJD precautions, and the occupational risks and hazards related to CJD.
- ✧ Single-use protective equipment such as gloves, gowns, waterproof aprons, masks, and protective eye wear⁽⁸⁵⁾ should be worn.
- ✧ The body should be placed on a plastic, impermeable sheet or body pouch so that suture site leakage can be contained.
- ✧ Perfusion drain sites should be similarly arranged to avoid surface exposure.
- ✧ All drainage fluids should be collected in a leak proof container.
- ✧ Perfusions and autopsy incisions sites should be closed with cyanoacrylates (super glue) and then wiped with bleach⁽¹⁾. The entire body should be wiped down with bleach⁽¹⁾.
- ✧ At the conclusion of the perfusion procedure, the container(s) of drainage fluids should be sealed, labelled “biohazardous”, and incinerated.
- ✧ The plastic sheet and other disposable material (e.g. exposed personal protective equipment) that have been in contact with bodily fluids or tissues should be sealed in a leak proof container, labelled “biohazardous”, and incinerated.
- ✧ After embalming, reusable instruments, equipment, and working surfaces in the preparation room (mortuary) should be thoroughly cleaned and decontaminated using the CJD decontamination processes listed in Table 6.

Category B; Grade III

Recommendations for Occupational Safety

Occupational health counselling should include the fact that no known cases of CJD have occurred through occupational accident or injury. A number of strategies to minimize the theoretical risk of infection following accidents have been proposed, but their usefulness is untested and unknown. Currently, the following common-sense actions are recommended⁽¹⁾.

- a. HCWs who come in contact with high risk or at risk patients should be educated about the epidemiology of CJD in order to have a clear understanding of the mode of transmission of the disease, what the risk factors are, and when CJD precautions are required^(12,13).

Category B; Grade III

- b. Appropriate single-use personal protective equipment^(11-13,106), such as gloves, gowns, masks and protective eye wear, should be worn by HCWs who are exposed to high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF of an at risk patient.

Category B; Grade III

- c. When there is accidental contamination of unbroken skin with high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF of an at risk patient, wash the skin with detergent and copious quantities of warm water (avoid scrubbing), rinse, and dry. Brief exposure (1 minute) to 0.1N NaOH or a 1:10 dilution of bleach can be considered for maximum safety⁽¹⁾. Report to Occupational Health⁽¹³⁾, according to procedures already in place.

Category B; Grade III

- d. For an accidental needlestick injury or laceration with equipment or an instrument involving high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF of an at risk patient, gently encourage bleeding; immediately wash the wound with copious warm soapy water (avoid scrubbing); rinse, dry, and cover with a waterproof dressing. Further treatment (e.g. sutures) should be appropriate to the type of injury⁽¹⁾. Report to Occupational Health⁽¹⁾, according to procedures already in place.

Category B; Grade III

- e. If splashing of high or low infectivity tissues of a high risk patient or high infectivity tissues and CSF from an at risk patient occurs into the eye or mouth, then immediately irrigate with either saline (eye) or tap water (mouth). Report to Occupational Health⁽¹³⁾, according to procedures already in place.

Category B; Grade III

- f. Health and safety guidelines call for mandatory reporting of injuries. Records should be kept for an extended period of time. (Note: The WHO Consultation on Infection Control Guidelines for Transmissible Spongiform Encephalopathies recommended that records should be kept for at least 20 years⁽¹⁾. The U.K. Advisory Committees on Dangerous Pathogens and Spongiform Encephalopathy recommended keeping a record of employees exposed to human TSE agents for 40 years after the last known exposure⁽¹²⁾).

Category B; Grade III

Recommendations for Protocol Failure

In the event that the established protocols are not followed, resulting in potential contamination and/or exposure to the CJD agent, a contingency protocol should be activated. Components of that protocol are listed in Part B - Table 4.

Category B; Grade III

Appendix I: Guideline Rating System

Categories for the strength of each recommendation	
Category	Definition
A	Good evidence to support a recommendation.
B	Moderate evidence to support a recommendation.
C	Insufficient evidence to support a recommendation.
Categories for the quality of evidence on which recommendations are made	
Grade	Definition
I	Evidence from at least one properly randomized, controlled trial.
II	Evidence from at least one well-designed clinical trial without randomization, from cohort or case-controlled analytic studies (preferably from more than one centre), from multiple time series, or from dramatic results in controlled experiments.
III	Evidence from opinions of respected authorities on the basis of clinical experience, descriptive studies or reports of expert committees.

Appendix II: Surveillance Definitions for Classic CJD⁽¹²⁵⁾

National Surveillance Case Definition for Classic Creutzfeldt-Jakob Disease⁽¹²⁶⁾

1. Sporadic Case

Confirmed CJD

- ✧ spongiform encephalopathy in cerebral and/or cerebellar cortex and/or subcortical grey matter
AND/OR
- ✧ encephalopathy with prion protein (PrP) immunoreactivity (plaque and/or diffuse synaptic and/or patchy/perivacuolar types)
AND/OR
- ✧ scrapie associated fibrils (SAF)

Probable CJD

- ✧ rapidly progressive dementia
AND
- ✧ typical EEG
AND
- ✧ at least two out of the following four clinical features: myoclonus, visual or cerebellar disturbances (ataxia), pyramidal/extrapyramidal dysfunction, akinetic mutism

OR

- ✧ rapidly progressive dementia
AND
- ✧ two out of the four clinical features listed above
AND
- ✧ duration of illness < 2 years
AND
- ✧ 14-3-3 positivity (in cerebrospinal fluid)

Possible CJD

- ✧ rapidly progressive dementia
 AND
- ✧ two out of the four clinical features listed above
 AND
- ✧ duration of illness < 2 years

2. Iatrogenic CJD

- ✧ progressive cerebellar syndrome in a pituitary hormone recipient
- ✧ sporadic CJD with a recognized exposure risk (e.g. dura mater transplant)

3. Familial CJD

- ✧ confirmed or probable sporadic CJD *plus* confirmed or probable CJD in a first degree relative
 AND/OR
- ✧ neuropsychiatric disorder *plus* disease-specific PrP mutation

4. Gerstmann-Sträussler-Scheinker (GSS)

- ✧ GSS in a family with dominantly inherited progressive ataxia
 AND/OR
- ✧ dementia and one of a variety of PrP gene mutations:
 - encephalo(myelo)pathy with multicentric PrP plaques

5. Familial Fatal Insomnia (FFI)

- ✧ FFI in a member of a family with PrP178 mutation:
 - thalamic degeneration, variable spongiform change in cerebrum

6. Kuru

- ✧ Kuru in the Fore population of Papua New Guinea:
 - although most neurologic features correspond to those of CJD with plaques, kuru should be diagnosed only in members of the Fore population in Papua New Guinea

Appendix III: Material Safety Data Sheets (MSDS)

The Canadian Centre for Occupational Health and Safety website is a source for MSDS:
<http://ccinfoweb.ccohs.ca/>

1. Example of portions of an MSDS submission for a sodium hydroxide preparation with manufacturer identifiers removed:

Section 1 - Chemical Product and Company Identification

Section 2 - Composition, Information on Ingredients

Section 3 - Hazards Identification - Emergency Overview

Corrosive Potential Health Effects

Eye:	Causes severe eye burns.
Skin:	Causes skin burns. May cause deep, penetrating ulcers of the skin.
Ingestion:	Causes gastrointestinal tract burns. Causes severe pain, nausea, vomiting, diarrhea, and shock.
Inhalation:	Irritation may lead to chemical pneumonitis and pulmonary edema. Causes severe irritation of upper respiratory tract with coughing, burns, breathing difficulty, and possible coma.
Chronic:	Prolonged or repeated skin contact may cause dermatitis.

Section 4 - First Aid Measures

- Eyes:** Immediately flush eyes with plenty of water for at least 15 minutes, occasionally lifting the upper and lower eyelids. Get medical aid immediately.
- Skin:** Get medical aid immediately. Immediately flush skin with plenty of soap and water for at least 15 minutes while removing contaminated clothing and shoes. Discard contaminated clothing in a manner which limits further exposure.
- Ingestion:** Do NOT induce vomiting. If victim is conscious and alert, give 2-4 cupfuls of milk or water. Never give anything by mouth to an unconscious person. Get medical aid immediately.
- Inhalation:** Remove from exposure area to fresh air immediately. If not breathing, give artificial respiration. If breathing is difficult, give oxygen. Get medical aid.
- Notes to Physician:

Section 5 - Fire Fighting Measures

- General Information:** Wear appropriate protective clothing to prevent contact with skin and eyes. Wear a self-contained breathing apparatus (SCBA) to prevent contact with thermal decomposition products. Use water with caution and in flooding amounts.
- Extinguishing Media:** For small fires, use dry chemical, carbon dioxide, water spray or alcohol-resistant foam.

Section 6 - Accidental Release Measures

- General Information:** Use proper personal protective equipment as indicated in Section 8.
- Spills/Leaks:** Absorb spill with inert material (e.g. vermiculite, sand or earth), then place in suitable container.

Section 7 - Handling and Storage

- Handling:** Wash thoroughly after handling. Use with adequate ventilation. Do not get in eyes, on skin, or on clothing. Do not ingest or inhale.
- Storage:** Store in a cool, dry, well-ventilated area away from incompatible substances. Keep away from strong acids. Keep away from metals. Keep away from flammable liquids. Keep away from organic halogens.

Section 8 - Exposure Controls, Personal Protection

Engineering Controls: Use adequate general or local exhaust ventilation to keep airborne concentrations below the permissible exposure limits.

Personal Protective Equipment

Eyes: Wear appropriate protective eyeglasses or chemical safety goggles as described by OSHA's eye and face protection regulations in 29 CFR 1910.133 or European Standard EN166.

Skin: Wear appropriate gloves to prevent skin exposure.

Clothing: Wear appropriate protective clothing to prevent skin exposure.

Respirators: Follow the OSHA respirator regulations found in 29CFR 1910.134 or European Standard EN 149. Always use a NIOSH or European Standard EN 149 approved respirator when necessary.

Section 9 - Physical and Chemical Properties

Section 10 - Stability and Reactivity

Chemical Stability: Stable.

Conditions to Avoid: Incompatible materials, acids.

Incompatibilities with Other Materials:

Reacts with mineral acids to form corresponding salts; reacts with weak acids gases like hydrogen sulfide, sulfur dioxide, and carbon dioxide; ignites when in contact with cinnamaldehyde or zinc; and reacts explosively with a mixture of chloroform and methane. Corrosive to metals such as aluminum, tin, and zinc as well as to alloys such as steel, and may cause formation of flammable hydrogen gas.

Hazardous Decomposition Products: Toxic fumes of sodium oxide, sodium peroxide fumes.

Hazardous Polymerization: Has not been reported

Section 11 - Toxicological Information

Section 12 - Ecological Information

Section 13 - Disposal Considerations

Dispose of in a manner consistent with federal, state, and local regulations.

Section 14 - Transport Information

Canadian TDG: No information available.

Section 15 - Regulatory Information

US Federal

TSCA: CAS# is listed on the TSCA inventory.
CAS# is listed on the TSCA inventory.
This material does not contain any Class 2 Ozone depletors.

Clean Water Act: CAS# is listed as a Hazardous Substance under the CWA.
None of the chemicals in this product are listed as Priority Pollutants under the CWA.
None of the chemicals in this product are listed as Toxic Pollutants under the CWA.

OSHA: None of the chemicals in this product are considered highly hazardous by OSHA.

State

European/International Regulations

European Labeling in Accordance with EC Directives

Hazard Symbols: Not available.

Risk Phrases:

Safety Phrases: Keep container tightly closed in a cool, well-ventilated place.
Do not inhale gas/fumes/vapour/spray.
Avoid contact with skin and eyes.
In case of contact with eyes, rinse immediately with plenty of water and seek medical advice.
After contact with skin, wash immediately with plenty of water.
Wear suitable protective clothing, gloves and eye/face protection.

WGK (Water Danger/Protection)

CAS#
CAS# No information available.

United Kingdom Occupational Exposure Limits

CAS# 1 OES-United Kingdom, STEL 2 mg/m³ STEL
CAS# OES-United Kingdom, STEL 2 mg/m³ STEL

Canada

CAS# is listed on Canada's DSL List.

CAS# is listed on Canada's DSL List.

CAS# is listed on Canada's Ingredient Disclosure List.

CAS# is not listed on Canada's Ingredient Disclosure List.

Exposure Limits

CAS#: OEL-AUSTRALIA:TWA 2 mg/m³

OEL-BELGIUM:STEL 2 mg/m³

OEL-DENMARK:TWA 2 mg/m³

OEL-FINLAND:TWA 2 mg/m³

OEL-FRANCE:TWA 2 mg/m³

OEL-GERMANY:TWA 2 mg/m³

OEL-JAPAN:STEL 2 mg/m³

OEL-THE NETHERLANDS:TWA 2 mg/m³

OEL-THE PHILIPPINES:TWA 2 mg/m³

OEL-SWEDEN:TWA 2 mg/m³

OEL-SWITZERLAND:TWA 2 mg/m³;STEL 4 mg/m³

OEL-THAILAND:TWA 2 mg/m³

OEL-TURKEY:TWA 2 mg/m³

OEL-UNITED KINGDOM:TWA 2 mg/m³;STEL 2 mg/m³

OEL IN BULGARIA, COLOMBIA, JORDAN, KOREA check ACGIH TLV

OEL IN NEW ZEALAND, SINGAPORE, VIETNAM check ACGI TLV

Section 16 - Additional Information

MSDS Creation Date: 9/02/1997 Revision #1 Date: 8/02/2000

The information above is believed to be accurate and represents the best information currently available to us. However, we make no warranty of merchantability or any other warranty, express or implied, with respect to such information, and we assume no liability resulting from its use. Users should make their own investigations to determine the suitability of the information for their particular purposes. In no way shall the company be liable for any claims, losses, or damages of any third party or for lost profits or any special, indirect, incidental, consequential or exemplary damages, howsoever arising, even if the company has been advised of the possibility of such damages.

2. Example of a portion of a sodium hypochlorite preparation MSDS submission with the manufacturer's identifiers removed

Identification

MSDS Record Number:
Product Name(S): SODIUM HYPOCHLORITE 5.25% Bleach
Date Of MSDS: 1997-08

Manufacturer Information

Manufacturer:
Address:
Emergency Telephone No.:

Material Safety Data

SODIUM HYPOCHLORITE 5.25% Bleach

Material Safety Data Sheet

Date issued: August 97

Product Identification

Product name: 5.25% bleach
Chemical name: Sodium hypochlorite
Product use: Bleaching agent and disinfectant

Hazardous Ingredients

ACGIH			
Hazardous Ingredient	%	TLV	CAS number
Sodium Hypochlorite	5.25	0.5 ppm (as Cl ₂)	

Physical Properties

Appearance and Odour: Clear greenish yellow liquid with chlorine odor
Boiling Point (deg C): Slowly decomposes at 40 deg C to NaCl and NaClO₃
Melting/Freezing Point: ~ - 5 deg C
Specific gravity: ~ 1.07 g/mL
Evaporation point: Not available
Solubility: Miscible in all proportions in water
pH: ~ 12.25

Reactivity Data

Stability: Unstable above 40 deg C, or in contact with metals or sunlight
Hazardous Polymerization: Will not occur
Materials to Avoid: Acids, ammonia, oxidizable materials, urea, most metals
Hazardous Decomposition or Combustion Products: Chlorine gas when in contact with acids.

Fire and Explosion Data

Flammability: No
Flash point: Not applicable
Autoignition Temperature: Not applicable
Fire Extinguishing media: Use appropriate media for surrounding fire.

Toxicological and Health Data

Route of entry: Skin contact, eye contact, inhalation and ingestion

Toxicological data:

Irritancy of material: Corrosive and irritant, risk of severe damages when eye contact

Effect of acute exposure: May cause discomfort, irritation, chemical burns

First Aid Measures

Inhalation: Move victim to fresh air. Obtain medical attention if necessary

Skin contact: Flush area with water for 15 minutes. Obtain medical attention if necessary

Eye contact: Flush eye with running water for at least 15 minutes. Obtain medical attention immediately.

Ingestion: Rinse mouth with water. Give ½ to 1 glass of water. Do not induce vomiting. Obtain medical attention immediately.

Preventives Measures

Respiratory protection: Not necessary if area is well ventilated

Skin protection: Rubber gloves are recommended

Eye protection: Safety glasses are recommended

Engineering controls: Local exhaust ventilation

Storage temperature (deg C): Between 0 deg C and 29 deg C, away from the sunlight

Handling procedures and equipment: Protect containers from damages. Avoid splashing.

Environmental Protection Data

Steps in the event of a spill or leak: Absorb and rinse with water small leaks.

Waste disposal methods: Consult federal, provincial and local regulations.

Appendix IV: Instrument Construction

The large majority of surgical instruments are manufactured from stainless steel. This can vary in quality (there are over 60 types of steel). Major European and U.S. manufacturers usually use high quality steel, but instruments of other origin may be made from lower grade steel, which is difficult to clean effectively.

The finish on an instrument can be polished or matt, and matt finished devices are more difficult to clean. Other materials, such as aluminum, titanium and plastics, can make up part or the whole of an instrument structure. Aluminum and plastic are more difficult to clean than high grade stainless steel. Titanium devices should clean easily. Construction of devices varies from simple “single surface” to complex, multi-jointed or multi-part construction.

The following categorization of instruments¹ may help in considering how easy a particular instrument might be to clean. Expert advice should be sought on instruments whose category is not clear.

Instrument category
<p>Category A: Can be decontaminated*</p> <ul style="list-style-type: none"> Single surface, no working parts Jointed smooth jaws and no ratchet Jointed with serrated jaws and ratchet Multi-part instrument that can be dismantled into component parts
<p>Category B: Varying degree of decontamination possible</p> <ul style="list-style-type: none"> Multi-part/jointed instruments that cannot be fully dismantled Instruments with lumen
<p>Category C: Impossible to guarantee safe decontamination**</p> <ul style="list-style-type: none"> Power tools (air or electric driven), not machine washable Exotic kit with multi-part, multi-material, only partly strippable Fibre optic flexible scopes Instruments with lumen

* If made from poor quality steel, instruments may not be effectively decontaminated.

** Some well-constructed kits in this category may be possible to decontaminate.

¹ Reproduced, with kind permission, from CJD Incidents Panel. *Management of possible exposure to CJD through medical procedures. A consultation paper.* Department of Health, United Kingdom, October 2001. www.doh.gov.uk/cjd/consultation

Appendix V: Example of Hospital CJD Risk Assessment and Management

Classic Creutzfeldt-Jakob Disease in Canada

Risk Assessment Tool

1. Use Table 2 to establish level of risk of patient

Table 2: Patient Risk for CJD ⁽¹⁾	
High Risk Patient	At Risk Patient*
<p>Diagnosed CJD**</p> <p>Suspected CJD:** undiagnosed, unusual, progressive neurological disease consistent with CJD** (e.g. dementia, myoclonus, ataxia, etc.)</p>	<p>Only under conditions in which there could be exposure to their high infectivity tissues, including CSF (see Table 3):</p> <p>Recipients of human dura mater grafts,[†] corneal grafts,[§] and human pituitary hormones</p> <p>Members of families with familial CJD, GSS and FFI[¶]</p>

* The incidence of CJD in Canada does not justify classifying people who have undergone neurosurgical procedures as at risk patients

** All forms of classic CJD (sporadic, familial and iatrogenic), including GSS and FFI, excluding variant CJD

† Recipients of dura mater grafts are frequently unaware of having received the graft.

§ Corneal grafts originating in a jurisdiction that requires the graft donor to be evaluated for neurological disease are not considered a risk for CJD⁽⁷⁶⁾.

¶ Familial CJD, GSS and FFI can be determined by genetic testing or may be indicated by the occurrence of two or more cases of CJD in the family (parent, child, sibling)⁽⁷⁶⁾.

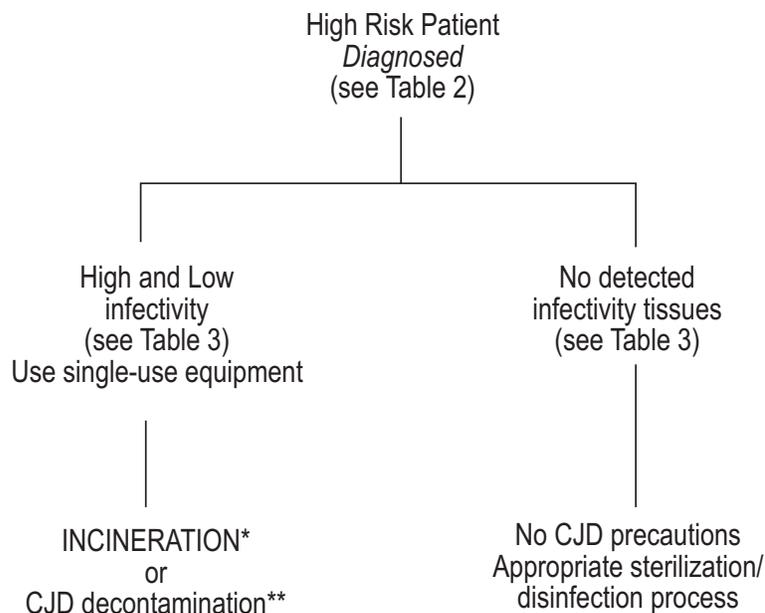
2. Use Table 3 to assess tissue risk

Table 3: Tissue Risk for CJD ⁽¹⁾	
Level of Infectivity	Tissues, Secretions and Excretions
High Infectivity	Brain, spinal cord, dura mater, pituitary, eye* (including optic nerve and retina)
Low Infectivity	CSF,** kidney, liver, lung, lymph nodes, spleen, placenta ⁽⁸⁸⁾
No Detected Infectivity	Adipose tissue, skin, adrenal gland, heart muscle, intestine, peripheral nerve, prostate, skeletal muscle, testis, thyroid gland, feces, milk, nasal mucus, saliva, serous exudate, sweat, tears, urine, blood, bone marrow, semen

* The highest levels of infectivity in the eye are associated with the optic nerve and retina and, to a much lower level, the cornea. It is expected that levels of infectivity for other parts of the eye are low or nonexistent. There is no infectivity with tears.

** Although CSF is classified as low infectivity and is less infectious than high infectivity tissues, it was felt that instruments contaminated by CSF should be handled in the same manner as those contacting high infectivity tissues in high risk and at risk patients⁽¹⁾.

Figure 1: Algorithm for the Management of Instruments and Equipment used on a High Risk Patient Diagnosed with CJD (Use single-use instruments and equipment wherever possible.)

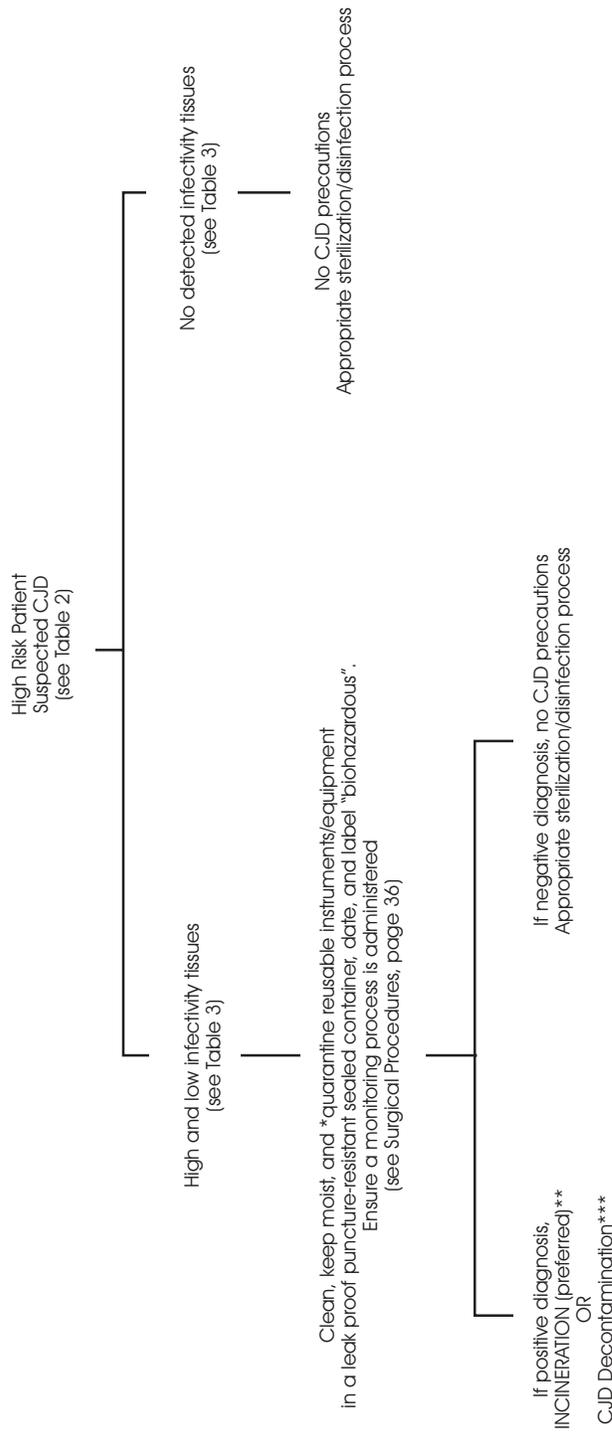


NOTE: For detailed information, refer to page 32: Recommendations for the management of a high risk or an at risk patient for CJD in the health care setting, in particular D. Medical Procedures and E. Surgical Procedures.

* Complex instruments, e.g. fiberoptic endoscopes, that cannot be adequately decontaminated by the chemical/sterilizer methods as per Table 6 should be incinerated (see Appendix IV).

** Reusable instruments and equipment should be critically assessed for dismantling (see Part B. Risk Management for CJD, Advance Planning, page 21).

Figure 2: Algorithm for the Management of Instruments and Equipment used on a High Risk Patient with Suspected CJD (Use single-use instruments and equipment wherever possible.)



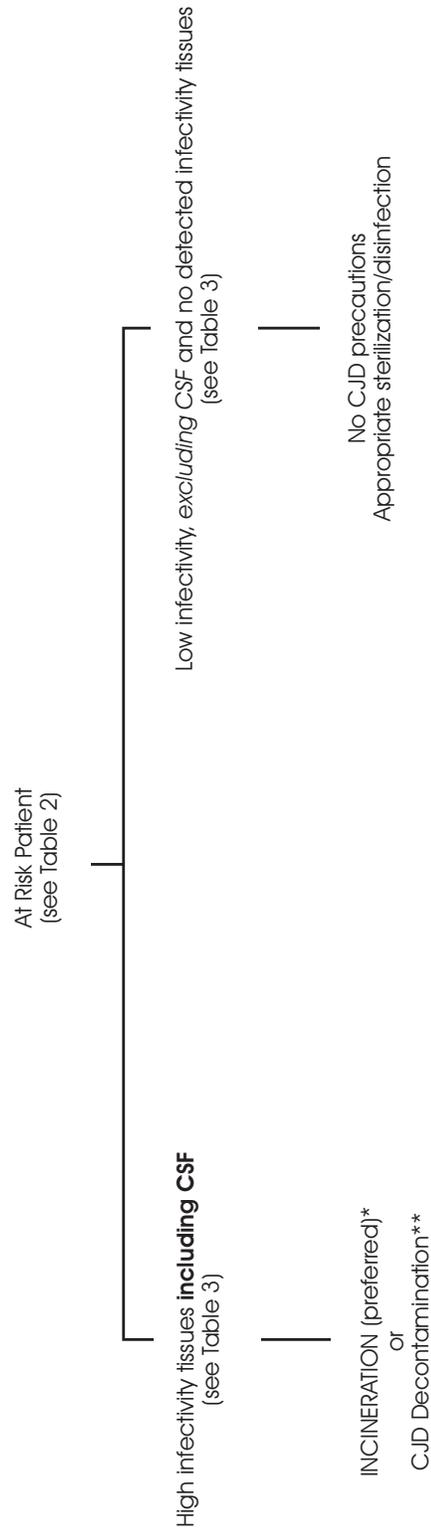
Note: For detailed information refer to page 32: Recommendations for the management of a high risk or an at risk patient for CJD in the health care setting, in particular D. Medical Procedures and E. Surgical Procedures.

* If reusable instruments are not quarantined from a high risk patient in whom CJD is suspected but not confirmed, then they must be treated as if used on a patient with diagnosed CJD (see Figure 1).

** Complex instruments, e.g. fiberoptic endoscopes, that cannot be adequately decontaminated by the chemical/sterilizer methods as per Table 6 should be incinerated. (see Appendix IV).

*** Reusable instruments or equipment should be critically assessed for dismantling (see Part B. Risk Management for CJD, Advance Planning, page 21).

Figure 3: Algorithm for the Management of Instruments and Equipment used on an At Risk Patient for CJD (Use single-use instruments and equipment wherever possible for high infectivity tissue and CSF.)



Note: For detailed information refer to page 32: Recommendations for the management of a high risk or an at risk patient for CJD in the health care setting, in particular D. Medical Procedures and E. Surgical Procedures.

* Complex instruments, e.g. fiberoptic endoscopes, that cannot be adequately decontaminated by the chemical/sterilizer methods as per Table 6 should be incinerated (see Appendix IV).

** Reusable instruments or equipment should be critically assessed for dismantling (see Part B. Risk Management for CJD, Management of Equipment and Environmental Surfaces, page 19).

Table 6: CJD Decontamination Processes⁽¹⁾

These recommendations are based on the best available evidence at this time and are in decreasing order of effectiveness. They must be followed, without exception, when there is exposure to high and low infectivity tissues from a high risk patient and high infectivity tissues and CSF from an at risk patient (see Tables 2 and 3). NOTE: If the instrument or surface cannot be fully immersed or flooded with the chemical disinfectant, then the item must be incinerated.

1. **Incineration:** Use for all instruments, effluent materials, and solid waste.

2. **Instrument Decontamination** for heat-resistant reusable instruments that an institution is unwilling or unable to incinerate.

2.1 **Cleaning:** Removal of adherent particles through mechanical or manual cleaning must be completed prior to any chemical/sterilizer decontamination of instruments. Instruments and other materials to be decontaminated should be kept moist between the time of exposure to infectious materials and subsequent decontamination; then

2.2 Immerse instruments in 1N sodium hydroxide (NaOH) or sodium hypochlorite* solution for 1 hour; remove from chemical solution; rinse instruments well; then immerse instruments in water and place in a sterilizer selecting the liquid cycle and heat to 121° C for 1 hour; or

2.3 Immerse instruments in 1N NaOH or sodium hypochlorite* for 1 hour; remove instruments from chemical solution; thoroughly rinse instruments in water; then transfer to an open pan, place in prevacuum sterilizer, and heat to 134° C for 1 hour or to 121° C in a gravity displacement sterilizer for 1 hour.

3. Hard Surface Decontamination

3.1 Remove visible soil.

3.2 Flood with 2N NaOH or undiluted sodium hypochlorite; let stand for 1 hour; then mop up and rinse with water; or

3.3 If surfaces cannot tolerate NaOH or undiluted sodium hypochlorite, thorough cleaning will remove most infectivity by dilution, and some additional benefit may be derived from the use of one or another of the partially effective methods listed in Table 5.

4. Chemical/Sterilizer Process for Dry Goods

4.1 Small dry goods that can withstand either NaOH or sodium hypochlorite should first be immersed in one or the other solution (as described in 2.3 above) and then heated in a prevacuum steam sterilizer to 134° C for 1 hour.

4.2 Bulky dry goods or dry goods of any size that cannot withstand exposure to NaOH or sodium hypochlorite should be heated in a prevacuum steam sterilizer to 134° C for 1 hour.

*20,000 ppm available chlorine

Note: Table 6 recommends that, when possible, two or more different methods of prion inactivation be combined sequentially in any decontamination procedure. Procedures that employ NaOH and heat consecutively appear to be sterilizing under worst-case conditions (e.g. infected brain tissue partly dried on the surfaces).

NaOH (or soda lye): Be familiar with and observe safety guidelines for working with NaOH (see MSDS sheet example in Appendix III). 1N NaOH is a solution of 40g NaOH in 1 litre of water. 1N NaOH readily reacts

with CO₂ in air to form carbonates that neutralize NaOH and diminish its disinfective properties. 10 NaOH solutions do not absorb CO₂, therefore 1N NaOH working solutions should be prepared fresh for each use either from solid NaOH pellets or by dilution of 10N NaOH stock solution⁽¹⁾. NaOH should be dissolved by adding the pellets carefully. Adding pellets to water may cause spitting, putting the individual preparing the solution at risk of injury. Do not use on aluminum.

Sodium hypochlorite (bleach): Be familiar with and observe safety guidelines for working with sodium hypochlorite (see MSDS sheet example in Appendix III). Household or industrial strength bleach is sold at different concentrations, so that a standard dilution cannot be specified. Its efficacy depends on the concentration of available chlorine and should be 20,000 ppm available chlorine. A common commercial formulation is 5.25% bleach with 52,500 ppm available chlorine⁽⁵⁾. Solutions should be prepared fresh for each use.

Reusable instruments or instrument components that an institution is unwilling or unable to incinerate must be reprocessed through one of the CJD decontamination processes listed in Table 6. For instruments not incinerated, after the CJD decontamination process the reusable instruments must be rechallenged through the appropriate decontamination process used for all reusable instruments (i.e. cleaned again, inspected, function tested, packed, and disinfected or sterilized before being made available for use on another patient).

Risk Assessment Tool/Pre-op Check List

To be completed for all neurosurgical/spinal surgeries by surgeon (with patient). This is done with the surgical consent form.

Risk assessment for identification of possible CJD patients includes the following questions:

1.	Does the patient have known/suspected CJD (established diagnosis)?*		
2.	Is there a family history of CJD?	Yes <input type="checkbox"/>	No <input type="checkbox"/>
3.	Is there a family history of any other inheritable spongiform encephalopathy (Gerstmann-Straussler-Scheinker or Fatal Familial Insomnia)?	Yes <input type="checkbox"/>	No <input type="checkbox"/>
4.	Has the patient ever received any human pituitary growth hormone therapy?	Yes <input type="checkbox"/>	No <input type="checkbox"/>
5.	Does the patient have a history of receiving any human dural engraftment?	Yes <input type="checkbox"/>	No <input type="checkbox"/>
6.	Was the patient investigated with depth electrode for epilepsy?	Yes <input type="checkbox"/>	No <input type="checkbox"/>
7.	Does the patient have a history of receiving a human corneal transplant?	Yes <input type="checkbox"/>	No <input type="checkbox"/>
8.	Does the patient have a rapidly progressive dementia not yet diagnosed?	Yes <input type="checkbox"/>	No <input type="checkbox"/>
9.	Has the patient received notification from their family physician that they have received blood products from a CJD donor?	Yes <input type="checkbox"/>	No <input type="checkbox"/>
	or		
	been in surgical contact with a possible CJD patient?	Yes <input type="checkbox"/>	No <input type="checkbox"/>
10.	Any other concerns?		

* When the quartet of dementia, myoclonus, periodic EEG activity and rapid progression is present, the diagnosis of CJD is almost certain.

Diseases that must be ruled out are Alzheimer's, Parkinson's, familial myoclonic dementia
One "Yes" answer requires further assessment by the surgeon and OR using CJD OR Guidelines (prior to surgery).

When high risk is suspected, notify the surgeon, OR, and Infection Control.

Date: _____ Physician Signature: _____

Patient Signature: _____

Source: Adapted with permission from the Hotel-Dieu Grace Hospital, Windsor, ON Canada

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