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Series on Infection Control

***Creutzfeldt-Jakob
Disease and Other
Human Transmissible
Spongiform
Encephalopathies***

***Guidelines on patient management
and infection control***

December 1995

Creutzfeldt-Jakob Disease and Other Human Transmissible Spongiform Encephalopathies

**Guidelines on patient management
and infection control**

This report was prepared by a Working Party
of the National Health Advisory Committee
of the National Health and Medical Research Council

National Health and Medical Research Council

NH M R C

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Glossary and abbreviations

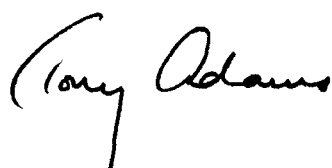
AS	Australian Standard; see Appendix 1
Asymptomatic	without symptoms
BSE	bovine spongiform encephalopathy – a transmissible scrapie disease of cattle
CJD	Creutzfeldt-Jakob disease
Codon	the unit of genetic code for the building blocks of proteins
CSF	cerebrospinal fluid
Dementia	a state of mental deterioration
Dura mater	outer-most of the three membranes covering the brain and spinal cord
EEG	electroencephalogram
EMG	electromyograph
Etiology	the study of the causes of diseases
Familial CJD	CJD occurring in more than one member of a family, usually associated with a genetic mutation (eg at Codon 200)
FFI	fatal familial insomnia – a genetically transmitted variant of CJD
Gonadotrophin	any hormone having a stimulating effect on the gonads. FSH and LH are two such hormones, secreted by the anterior pituitary
GSS	Gerstmann-Striussler-Scheinkersyndrome – a familial type of CJD with genetic mutations
Homozygous	having identical alleles for a given gene
Iatrogenic	induced in a patient as a result of medical treatment
Kuru	a disease related to CJD, which occurred in epidemic form in a small group of people in Papua New Guinea
Neuro-degenerative	degeneration or decay of the nervous system
Parenteral	route of administration other than through the digestive tract
Pituitary hormones	hormones derived from pituitary gland eg growth hormone (GH) and follicular stimulating hormone (FSH)
Prion	an acronym for ‘ <u>I</u> nfectious <u>P</u> rotein’
PrP	prion protein or protease resistant protein
Proteolysis	the cleavage of proteins by enzymes
Scrapie	transmissible spongiform encephalopathy in sheep and goats
Sporadic CJD	CJD that occurs at random in the population
Standard Precautions	this term replaces ‘universal precautions’. Work practices which require everyone to assume that all blood and body substances are a potential sources of infection, independent of perceived risk. Such precautions involve the use of safe work practices and protective barriers.
Zoonotic transmission	transmitted to humans from animals

Foreword

The document '*Creutzfeldt-Jakob Disease and Other Human Transmissible Spongiform Encephalopathies: Guidelines for Patient Management and Infection Control*' was prepared by a working party convened by the National Health and Medical Research Council (NHMRC) to develop infection control guidelines for the management of, and for medical and surgical procedures including autopsies on, patients with or at risk of developing Creutzfeldt-Jakob disease (CJD).

The unique nature of the infectious agent of CJD, particularly its resistance to routine autoclaving and chemical disinfection, necessitated the formulation of disease-specific infection control procedures.

Experts from areas including infection control, neurosurgery, pathology, ophthalmology and dentistry contributed to the formulation of the guidelines, and I am confident that their contributions will assist health care workers and home carers to implement appropriate precautions, thereby reducing the risk of transmission of CJD.



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15 December 1995

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1. Introduction

Creutzfeldt-Jakob disease (CJD) is a rare degenerative disorder of the central nervous system which occurs sporadically at an annual rate of about one case per million people. It is one of several fatal neurodegenerative diseases (also known as transmissible spongiform encephalopathies) occurring in humans and animal species, and is characterised by microscopic vacuoles in the brain, astrocytosis and loss of neurones.

This group of diseases includes:

In humans:

Creutzfeldt-Jakob disease (CJD);
Gerstmann-Straussler-Scheinker syndrome (GSS);
kuru;
and fatal familial insomnia (FFI);

and in animals:

scrapie in sheep and goats;
bovine spongiform encephalopathy (BSE) in cattle;
feline spongiform encephalopathy in domestic cats;
transmissible mink encephalopathy (TME);
chronic wasting disease in captive mule deer; and
spongiform encephalopathy in captive exotic ungulates.

In 1968, it was discovered that CJD was infectious and could be transmitted by inoculating diseased brain tissue into the brains of non-human primates. At first, it was thought that the infectious agent was a virus or virus-like particle; however, in the 1980's it became clear that a normal host protein, called PrP, was an important component of the infectious agent. The significance of these studies increased when genetic mutations in the gene which codes for PrP were shown to be associated with the human disease.

It now appears that a modified form of PrP protein is the major, if not sole, component of the infectious agent. The nature of this modification appears to lie in its shape – an abnormally folded form of the PrP protein makes it very stable and resistant to proteolysis. Replication of infectivity is thought to be caused by one abnormal molecule inducing another molecule (the normal PrP found in most cells of the body) to assume a similar conformation.

The accumulation of protease-resistant PrP molecules in the brain, occurring over a long time interval, appears to be the basis of the neural degeneration and the clinical signs associated with these diseases. Experimental animals which lack the PrP gene are totally resistant to infection, which supports the concept that conformational changes in the normal PrP protein contribute to the disease state. The novel mechanism that a protein by itself can become infectious and self-replicating has led to the categorisation of CJD and other related disorders as the 'Prion' diseases (an acronym for Infectious Protein).

The basic epidemiological features of the disease are reviewed elsewhere^{11,27,46,47}. Approximately 15 per cent of all patients occur with familial clustering (this includes two related diseases, GSS and FFI) and in most of these a pathogenic mutation of the PrP gene can be demonstrated. In the remaining 85 per cent there is increasing evidence that the proportion with homozygosity at codon 129 of the PrP gene may be higher than in a normal population. Since this phenomenon may confer susceptibility to iatrogenic transmission, the possibility is raised that a significant proportion of 'sporadic' CJD may fall into this etiologic category.

In most instances of sporadic CJD, a thorough analysis of the patient's past medical and occupational histories fails to disclose clear evidence of an external source or point of infection. In a few patients however, there is a clear instance of iatrogenic contamination. Documented examples include dura mater grafts^{32,61} and one corneal graft, neurosurgical procedures^{36,65} and contaminated hormones (growth hormone; gonadotrophins) prepared from pituitaries post mortem^{3,34}. These instances of iatrogenic transmission demonstrate that short incubation periods (one to two years) are to be expected after direct intracerebral contamination, whereas very long incubation periods (one to two decades or more) occur after low dose peripheral contamination.

Although iatrogenic transmission of CJD is rare, health workers should be aware of the potential for transmission, and actively seek to implement suitable infection control procedures to minimise the risk of cross-contamination. The infectious agent is relatively stable in physico-chemical conditions that inactivate conventional viruses and bacteria, so it may persist in a viable form on instruments which have been autoclaved or decontaminated by normal chemical procedures. Once a patient is infected, there may be an incubation period ranging from two to forty years before the onset of disease. During this time the abnormal accumulation of PrP does not elicit any immune/inflammatory response and currently no blood test is available to determine if infection has occurred. Examination of brain tissue by biopsy or autopsy remains the only available method for obtaining a definitive diagnosis.

Formulation of the following guidelines involve consideration of several important factors:

- the unusual resilience of the PrP protein;
- patient risk status;
- distribution of infectious agent in various tissues and body fluids; and
- similarities between CJD and other related animal diseases from which knowledge/experience has been gained.

It is hoped that these guidelines will assist health professionals in implementing more stringent infection controls, which may prevent the inadvertent transmission of CJD.

2. Transmission of CJD

2.1 Routes of transmission

Within a highly susceptible species, direct intracerebral inoculation of relatively low doses of infectious material will yield close to 100 per cent 'takes' or successful transmissions. In contrast, in the same species, even high doses given by a peripheral route (eg subcutaneous, conjunctival or oral) will yield only a partially successful rate of transmission⁵³. The oral route of transmission is particularly inefficient, except where direct breaches of the oral mucosa occur²¹. Experimental studies have shown that for an infectious inoculum given peripherally, replication of infectivity begins first in the spleen, and then spreads to the central nervous system via the peripheral nervous system. Thus some mechanism of retrograde and trans-synaptic neural spread is involved.

Table 1 provides a guide to the relative distribution of infectivity that might be expected in different tissues/body fluids of a patient with CJD. This information is based on studies of experimentally transmitted CJD in non-human primates, and from other animal models. Experience to date demonstrates that the highest risk of iatrogenic transmission occurs when the central nervous system of persons with CJD is exposed. Particular attention is therefore required in the practice of neurosurgery, ophthalmic surgery, neurology (lumbar punctures) and in laboratory procedures in which brain tissue and CSF (cerebrospinal fluid) are exposed.

2.2 Vertical and lateral transmission

As CJD is such a rare disease it is difficult to assemble accurate epidemiological data. Nevertheless, studies of kuru (a related disease which occurred as an epidemic in the Eastern Highlands of Papua New Guinea) indicated that vertical transmission and lateral transmission (eg to health workers or others in contact with kuru patients) did not occur.

Studies of scrapie transmission in sheep has indicated that placenta may transmit the disease, therefore it is recommended that placenta should not be used in the preparation of pharmaceuticals.

There is no evidence that CJD can be transmitted through normal social contact.

2.3 Iatrogenic transmission

Iatrogenic transmission of CJD has occurred via corneal graft, dura mater grafts, neurosurgical instruments and from CJD contaminated human pituitary hormones. Some investigators have demonstrated infectivity^{44,45,54} and abnormal forms of the PrP molecule in blood, and it therefore appears prudent to treat blood as potentially infectious, even though there is no epidemiological evidence that blood transfusion is a major risk factor for CJD^{33,38}. Unsubstantiated claims of transfusion related CJD in Australia are being investigated⁴⁰.

2.4 Occupational transmission

Increased numbers of CJD patients who are health professionals^{8,9,48,51,62} or farmers exposed to bovine spongiform encephalopathies^{29,52} are being reported, although statistically it is not clear that they are over represented in the general CJD population. Similarly, it is still not clear whether previous surgical procedures are risk factors in the development of CJD²⁸.

2.5 Zoonotic risk of transmission

To date, there is no convincing evidence that scrapie and related diseases of animals can be transmitted to humans. However, when considering issues of public health and infection control, it is prudent not to overlook this possibility.

Scrapie in sheep and goats is common in Europe and North America. A similar disease, bovine spongiform encephalopathy, has occurred in cattle primarily in the United Kingdom. Fortunately, Australia is free from these animal diseases, but the inadvertent importation of contaminated products from the US or Europe is certainly a possibility. These products may include surgical catgut (made from sheep intestines), bone implants, cardiac patches, laboratory cell culture media (particularly bovine serum), some pharmaceuticals, some cosmetics and processed foods²⁵. Where direct parenteral contamination is possible (eg surgical catgut or pharmaceuticals), use of these products should be avoided.

Scrapie has occurred in epizootic fashion over the last two hundred years. Scrapie has been implicated as the causative agent of recent outbreaks of bovine spongiform encephalopathy in the UK²⁵. This epidemic appears to have arisen from the contamination of the food cycle with scrapie-infected carcasses. With the removal of this source of contamination, the epidemic is slowly coming under control.

These epidemics have important lessons for human control guidelines: CJD is normally a disease of low infectivity/transmissibility; however, under certain conditions of environmental contamination, a cycle of high infectivity/transmissibility can be created, but this cycle can be broken if appropriate measures are taken.

Table 1: Estimated levels of infectivity in tissues/body fluids

Tissue/body fluids	Estimated level of infectivity
Brain, spinal cord, eye (cornea), pituitary, dura mater	<p>Highest levels of infectivity</p> <p>The CNS, in areas affected by spongiform change, may contain levels of infectivity averaging 10^5 units/gm¹⁵. Maximum containment and decontamination procedures should be used for all patients when these tissues are exposed.</p>
Cerebrospinal fluid	<p>High levels of infectivity reported</p> <p>Maximum decontamination measures apply for all risk groups when handling CSF.</p>
Spleen, lymph nodes, tonsil, adrenal glands, lung, liver, kidney, thymus, placenta and membranes, peripheral nerve and pancreas	<p>Medium to low levels of infectivity</p> <p>Organs and tissues outside the CNS may demonstrate infectivity but at much lower levels. Maximum infection controls are therefore essential for Group 1 patients (see Section 4).</p>
Teeth and gingiva	<p>Unknown but treat as infectious</p> <p>There is uncertainty as to whether some dental procedures where vascular tissue is involved pose a risk of transmissibility of CJD. Dental procedures in which blood and/or gingival tissue are involved should therefore be regarded as a potential risk, particularly in Group 1 patients (see Section 4).</p>
Bone marrow, leukocytes, red blood cells and serum	<p>Probably very low or negligible infectivity</p> <p>There is still doubt as to whether blood components carry the infectious agent. Some investigators have claimed to have demonstrated infectivity^{44,45,54} and abnormal forms of the PrP molecule in blood. Although there is no epidemiological evidence to support blood transfusion as a major risk factor for CJD^{33,38}, blood should be treated as potentially infectious, particularly in Group 1 (high risk) patients (see Section 4).</p>
Tears, saliva, sputum, faeces, urine, semen, other bodily secretions and milk	<p>No reported presence of infectivity</p> <p>Most external bodily secretions have not shown infectivity, both in humans and in experimental animals¹⁵.</p>
Hair and skin	<p>Undetectable levels of infectivity</p> <p>There is no evidence to suggest that CJD can be spread by contact with intact skin (epidermis) or from hair and therefore normal infection controls should be adequate. However, when open wounds eg bedsores, abrasions or weeping rashes are present, particularly with Group 1 patients, maximum infection control procedures should be followed.</p>
Heart, skeletal muscle, cartilaginous tissue, connective tissue, adipose tissue, uterus and testis	<p>Low levels of infectivity or no reported presence of infectivity</p> <p>Although there have been no reports of detectable levels of infectivity in these tissues, maximum infection controls still apply for Group 1 patients because of the involvement of blood in most procedures involving these tissues.</p>

3. Diagnosis of Creutzfeldt-Jakob disease

Diagnosis of CJD is by clinical and neuropathological examination. The presence of triphasic sharp waves on the electroencephalogram is often of assistance in making the diagnosis. Imaging techniques are useful in excluding other causes of subacute dementia. Similarly, a lumbar puncture is often required to exclude other disease processes. Brain biopsy is sometimes indicated, and may present the only opportunity to make a definitive diagnosis if the prospects of obtaining an autopsy are not favourable.

The pathologic criterion of spongiform change (a diffuse vacuolation of the dendrites of neurons), together with neuronal loss and gliosis, is the best indicator of transmissibility of disease. Whether at biopsy or autopsy, if the immunocytochemical examination of brain tissue also demonstrates the accumulation of amyloid filaments composed of the PrP molecule, then a definitive diagnosis can be made (but such amyloid deposits are seen in less than 20 per cent of cases). In the future, diagnostic genetic, biochemical or quantitative immunochemical assays for the altered infection specific PrP protein may become available for use on brain tissue¹², CSF or peripheral blood monocytes. Such assays are not currently available in clinical laboratories.

The accuracy of clinical diagnosis is probably about 80 per cent, with a slightly higher rate in the pathological laboratory. This means that a certain proportion of false positive and negative cases will occur in routine clinical practice, a fact that needs to be taken into account when infection control guidelines are being implemented.

4. Patient risk status^{5,49,63,64}

From a practical point of view, it is convenient to divide the population at risk of spreading CJD into two groups, the rationale being that higher concentrations of infectivity are likely to be present in individuals grouped as high risk. More stringent infection controls are therefore needed when decontaminating instruments or performing medical procedures.

Group 1 : High-risk patients

This group includes:

- patients with proven CJD;
- patients with clinically suspected CJD;
- asymptomatic carriers of pathogenic mutations of PrP occurring within the context of familial CJD/GSS/FFI; and
- all members of a family with familial CJD/GSS/FFI in whom the PrP genotype is undetermined or uncertain⁵.

Group 2: Low-risk patients

This group includes:

- any patient with undiagnosed progressive neurological illness with or without dementia;
- all members of a family in which there is a strong family history of undiagnosed dementing/neurological illness;
- recipients of human pituitary hormones (growth hormone and gonadotrophins); and
- recipients of dura mater grafts or persons with a record of transdural neurosurgical operation between 1972 and 1989 (since 1987, the sourcing of dura mater for grafting has been more rigidly controlled and neurosurgeons have found substitutes for dural homografts).

Obviously if it can be ascertained by consultation with the operating neurosurgeon that dura mater grafting was not used in a particular individual, then that person would not fall into this Group 2 category.

5. Responsibilities of all health workers

It is the responsibility of all health workers:

- to be aware of the potential risks posed by patients in both CJD risk groups;
- to ensure that adequate decontamination procedures are implemented to minimise the risk of transmission of CJD to other patients, themselves and co-workers;
- to ensure that staff have adequate training and facilities to enable medical procedures to be carried out with minimum risk of transmission; and
- to ensure that equipment is maintained in sound working order and is subject to regular quality checks (eg steam sterilisation should comply with AS 4187; and corroded instruments which may not be effectively decontaminated should be replaced).

6. Responsibilities of people at risk of CJD

Patients (or an individual responsible for caring for a person with dementia) are advised to notify their doctors, dentists and other health workers of their status so that appropriate infection containment can be arranged.

This is particularly important for Group 1 (high risk) individuals (see below). As with other infections (eg HIV or Hepatitis B), there is an obligation to avoid passing the infectious agent to others".

7. Decontamination procedures^{2,4, 36, 63,64}

Whenever possible, objects and surfaces which are contaminated with potentially infectious material should be disinfected with heat and/or chemicals. Although no method currently available can guarantee complete sterilisation^{16,60}, the methods in Table 2 are those currently believed to be most effective in decreasing the level of infectivity.

Attention to the containment and to preparatory methods of decontamination are just as important as the processes themselves. Objects loaded into autoclaves must be arranged so that the steam has access to all potentially contaminated surfaces. Prior soaking of contaminated objects in a chemical disinfectant (such as sodium hydroxide), followed by washes in running tap water to reduce the amount of particulate material, would also be advisable.

All needles and other sharp objects should be placed in ridged puncture-proof sharps container (in accordance with AS 4031-1992) prior to being sent for disposal by incineration (see note page 10). Other materials sent for disposal should be enclosed within yellow infectious waste bags labelled with an international biohazard symbol and the words 'Medical Waste'.

It should be noted that the following decontamination procedures **are not adequate** for completely inactivating infectivity: UV or gamma irradiation, normal autoclaving (121°C at 15 psi or 101 kPa), glutaraldehyde and other aldehydes, boiling, dry heat sterilisation, ethylene oxide, acetone and alcohols.

Table 2: Recommended decontamination methods

Method	Application
<p>1. Autoclaving^{16,17,18,26,39,43,55,57}</p> <ul style="list-style-type: none"> - gravity displacement or porous load (prevacuum) autoclave** - 134°C (pressure is 30 psi or 203 kPa at 134°C) - holding time at temperature should be at least 18 minutes single cycle or six separate 3 minute cycles - loading must ensure that steam has access to all contaminated surfaces. 	<p>This method is suitable for all instruments that withstand autoclaving at this temperature/pressure. Where this is not the case, the other methods should be considered.</p> <p>** gravity displacement autoclaving may be less effective than porous load (prevacuum) autoclaving.</p> <p>Some authorities advise holding time at temperature for at least one hour^{20a}.</p>
<p>2. Exposure to 1–2 M sodium hydroxide^{17,18,58}</p> <p>Instruments:</p> <ul style="list-style-type: none"> - 1M NaOH equals 40 g NaOH per one litre of water - expose instruments to this solution for at least one hour at room temperature - make certain that instruments are completely submerged - physically scrub items to remove excess tissue/blood while in NaOH solution - make certain that all articles used for cleaning instruments are adequately decontaminated. <p>Surfaces:</p> <ul style="list-style-type: none"> - drench surface with NaOH solution - leave for at least one hour - use paper towels to absorb liquid - place paper towels directly into plastic bag and seal securely - dispose by incineration or other appropriate method of disposal. 	<p>This method is suitable for instruments that can not be autoclaved at the high temperatures required in method 1 (above).</p> <p>Caution Not suitable for some instruments (eg aluminium) as they may corrode.</p> <p>NaOH (sodium hydroxide) is caustic soda. Avoid contact with eyes and mucous membranes.</p> <p>1–2 M NaOH is the preferred method for cleaning contaminated surfaces (eg floors, benches splattered with blood or CSF).</p>
<p>3. Autoclaving as above but submerged in 1M NaOH or 3 per cent sodium dodecyl sulphate treatment.</p>	<p>This method may add an extra level of safety. It is only suitable for instruments that will withstand such harsh treatment.</p>
<p>4. Soaking in sodium hypochlorite^{13,18,39,57}</p> <ul style="list-style-type: none"> - hypochlorite solution should contain 2.0 to 2.5 per cent sodium hypochlorite - solution should be freshly prepared - soak instruments or surface for at least one hour - clean surfaces with paper towels following soaking and dispose by incineration as above. 	<p>This method is less effective than 1–2 M NaOH and is also less effective when used on surfaces where corrosion has occurred^{26,59}.</p> <p>Repeated use will cause corrosion of some metallic surfaces.</p>
<p>5. Other chemical treatments</p> <p>Formic acid (96 per cent)²⁰</p> <p>This chemical is particularly suitable for use in the pathology laboratory:</p> <ul style="list-style-type: none"> - fix small blocks of tissue in 4 per cent formaldehyde solution (10 per cent formol-saline) - follow by immersion in formic acid (> 96 per cent) for one hour - for machine processing, re-wash blocks in formalin as formic acid may damage plastic containers - where tissues are processed by hand, transfer directly from formic acid into ascending alcohol solutions. 	<p>Suitable for histology/pathology laboratory. This has been suggested as a suitable treatment when treating tissue sections. Other commonly used fixatives do not inactivate infectivity and therefore may pose a risk to laboratory staff during sectioning.</p> <p>Caution Formic acid is dangerous when in contact with skin, eyes etc.</p>

continued over page

Table 2: continued

Method	Application
<p>6. Incineration*</p> <ul style="list-style-type: none"> - immediately place soiled articles into a suitable container eg yellow infectious waste bag with international biohazard symbol and the words 'Medical Waste'. - incinerate as soon as possible adhering to normal infectious disease disposal procedures. - needles, blades and other sharp articles should be placed in a containers (in accordance with AS 4031-1992) prior to incineration. 	<p>Suitable for hospital and office practice waste including; swabs, wound dressings, needles, catheter tubing, single use protective clothing, and other single use equipment from surgical/other procedures involving treatment of Group 1 and 2 patients.</p> <p>Also suitable for linen soiled with CSF from hospital wards handling Group 1 patients.</p> <p>Also suitable for disposing of contaminated organs and tissue sections.</p>

- * In the absence of an Australian Standard, disposal of biological waste infected with CJD is to be in accordance with the best practice or equivalent standards prescribed under the laws, procedures, Codes of Practice or other regulatory provisions in force in the relevant State or Territory that are most consistent with this Guideline.

8. Injury resulting in contamination

8.1 Accidental contamination of unbroken skin

Where there is accidental contamination of unbroken skin with blood or body fluids from Group 1 (high risk) patient:

- wash skin using detergent and copious quantities of warm water;
- avoid vigorous scrubbing; and
- report the injury according to normal procedures for your organisation.

8.2 Accidental needle stick or lacerations

If these injuries occur when caring for CJD patients in risk Group 1 (high risk):

- wash wound immediately with copious amounts of warm water"; and
- report the injury according to normal procedures for your organisation.

8.3 Splashes into eye

If splashing of blood or CSF into the eye occurs when caring for Group 1 patients:

- immediately institute normal eye washing procedures using warm water; and
- report the incident according to normal procedures for your organisation.

9. General infection control guidelines^{2,4,36,49,63,64,66}

The Standard Precautions which apply to the prevention of blood borne pathogens such as Hepatitis B and HIV should apply to the management of patients with known or suspected CJD and related diseases. Health professionals, allied health workers, infection control and laboratory personnel and others who come into contact with CJD patients should be given appropriate training in infection control procedures. Because the clinical disease is sometimes difficult to diagnose and because of the known long incubation (preclinical) period, Standard Precautions and body substance isolation should apply in a wider context to all persons with undiagnosed progressive neurological disease, and to instruments and persons coming into direct contact with tissue related to the central nervous system.

Protective clothing should be worn routinely and skin wounds, eczematous lesions etc covered with waterproof dressings. If splashing is possible then protective eye-wear and masks should also be worn.

To prevent injury and possibility of infection for carers, sharp articles such as knives, scissors and glassware should be avoided. Where dispersal of material is necessary (eg mixing or centrifugation), sealed containers and closed centrifuge buckets are preferable and use of biological safety cabinets is recommended.

If there is accidental contamination of unbroken skin with blood or body fluids from a patient, wash the skin with detergent and copious quantities of warm water avoiding vigorous scrubbing. For accidental needle stick injuries or lacerations with equipment used on Group 1 (high risk) CJD patients, wash the wound immediately with copious quantities of warm water². Such injuries should be reported to relevant authorities at the institution. Again, it should be emphasised that no case of CJD infection arising as a result of this type of contamination has ever been recorded.

Whenever possible, objects and surfaces which are potentially contaminated with infectious material should be disinfected with heat and/or chemicals, although no method currently available can guarantee complete decontamination^{16,60}. Recommended methods of decontamination are presented in Table 2.

Contaminated single use equipment, linen and such-like should be incinerated, as should all needles and sharps. The use of yellow infectious waste bags with the international biohazard symbol and the words 'Medical Waste' and ridged puncture proof containers is advised for containment of waste material prior to incineration.

In dealing with contaminated surfaces, washing with 1–2 M sodium hydroxide is recommended. We suggest that surfaces be exposed to this chemical for at least one hour ie wipe, leave one hour, then clean up.

10. Specific infection control procedures

10.1 Neurosurgery and neuroradiology

The performance of neurosurgical and interventional neuroradiological procedures on patients in Group 1 is not contra-indicated, but obviously special attention is required. In addition to the Standard Precautions several specific recommendations are made which are consistent with those currently in place in the United

Patients who fall into Group 2 should be managed as routine, with particular attention given to decontamination of surgical instruments.

This committee recommends that the use of dura mater grafts of human origin cease unless the donor can be verified free of CJD.

Table 3: Neurosurgery and neuroradiology

Specific recommendations	
Group 1 (high risk)	Group 2 (low risk)
Standard Precautions	
Adhere to Standard Precautions for all procedures, in addition to extra precautions set out below.	
Use of instruments	
Essential to maintain one way flow of instruments for all procedures. Use single use equipment wherever possible.	Essential to maintain one way flow of instruments for all procedures. Use single uses in preference to re-usable equipment
Scheduling of patients	
Schedule operation or procedure at end of day to allow adequate cleaning of facilities.	End of day scheduling may be recommended depending on the procedure and the tissues involved.
Training and personnel	
Personnel must be aware of CJD risks and trained in adequate infection control procedures. Least number of people should participate in the operation/procedure.	Personnel must be aware of CJD risks and trained in adequate infection control procedures.
Protective clothing	
It is essential to wear protective clothing at all times. Use of single use protective clothing (eg gowns, caps) is advised.	Advisable to wear protective clothing at all times. Use of single use protective clothing is advised.

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Table 3: continued

Specific recommendations	
Group 1 (high risk)	Group 2 (low risk)
Disposal of contaminated protective clothing	
<p>Single use gowns etc should be sealed in yellow infectious waste bags with international biohazard symbol and the words 'Medical Waste' for disposal by incineration.</p> <p>Non-disposable protective clothing that has been soiled with CSF, brain tissue, neural tissue or blood should also be incinerated.</p>	<p>Incinerate single use clothing.</p> <p>Normal laundering and autoclaving is suitable for non-disposable gowns which are not soiled with blood, CSF, brain or neural tissue.</p> <p>Soiled (blood, CSF, brain or neural tissue) protective clothing should be incinerated.</p>
Reusable equipment	
<p>When it is necessary to use instruments not normally regarded as single use, these instruments should not be re-used and should be destroyed by incineration.</p>	<p>Continual reuse of reusable equipment is permitted only if adequate decontamination can be assured. eg minimum of 18 minutes autoclaving at 134 °C or 1 hour soaking in 1–2 M NaOH.</p>
Collection of specimens	
<p>Collect into a secure closing container and enclose in a plastic bag. The container must be labelled clearly with patient details, including a CJD risk alert to laboratory/ other medical personnel.</p>	<p>Standard Precautions apply.</p>
Disposal of specimens	
<p>Disposal of all specimens should be by incineration.</p>	<p>Brain and CSF samples should be incinerated.</p> <p>Disposal of other specimens should follow Standard Precautions.</p>
Other articles used in procedures	
<p>Use of reusable articles is not permitted. Swabs, dressings, linen, needles etc used during operations must be disposed of by incineration.</p>	<p>All articles coming into contact with CSF must be enclosed in yellow infectious waste bags with international biohazard symbol and the words 'Medical Waste' and disposed of by incineration.</p> <p>Standard Precautions apply for disposal of other waste materials.</p>

10.2 Neurology services

Since neurologists often have the primary responsibility for the initial diagnosis and management of CJD, the probability of contamination of instruments, in particular EMG needles, EEG electrodes, sensory testing pins and lumbar puncture needles needs to be borne in mind. For Group 1 and 2 patients, contaminated instruments should be treated as for neurosurgical instruments (see Table 3).

10.3 Interventional radiology, general surgery, ophthalmology and anaesthetics⁴²

For Group 1 patients, surgical instruments in contact with tissues outside the central nervous system still carry a substantial risk of contamination with infectivity. Non-disposable reusable equipment should therefore be discarded.

Persons in Group 2 should advise the treating surgeon/anaesthetist who will ensure that proper decontamination of instruments is performed.

Table 4: Interventional radiology, general surgery, ophthalmology and anaesthetics

Specific recommendations	
Group 1 (high risk)	Group 2 (low risk)
Standard Precautions	
Adhere to Standard Precautions for all procedures, in addition to extra precautions set out below.	
Scheduling of patients	
Schedule operation or procedure at end of day to allow adequate cleaning of facilities.	End of day scheduling may be recommended depending on the procedure and the tissues involved.
Training and personnel	
Personnel must be aware of CJD risks and trained in adequate infection control procedures. Least number of people should participate in the operation/procedure.	Personnel must be aware of CJD risks and trained in adequate infection control procedures.
Protective clothing	
It is essential to wear protective clothing at all times. Use of single use protective clothing (eg gowns, caps) is advised.	Advisable to wear protective clothing at all times.
Disposal of contaminated protective clothing	
Single use gowns etc should be sealed in yellow infectious waste bags with international biohazard symbol and the words 'Medical Waste' for disposal by incineration. Non-disposable protective clothing that has been soiled with CSF, brain tissue, neural tissue or blood should also be incinerated.	Incinerate single use clothing. Normal laundering and autoclaving is suitable for non-disposable gowns which are not soiled with neural tissue, CSF or large volumes of blood. Soiled (CSF, brain or neural tissue, or blood) protective clothing should be incinerated.
Surgical instruments	
Use single use equipment wherever possible. Instruments soiled with neural tissue, CSF or blood should be discarded, even if the instruments are normally reusable.	Use single use in preference to reusable equipment.
Reusable equipment	
When it is necessary to use instruments not normally regarded as single use: Tonometers which come into direct contact with corneas should be discarded after use on a Group 1 patient ³⁵ . Plastic tonometer covers may be used and then discarded. Non contact air or puff tonometers which totally avoid contact with the cornea should be used. Endoscopes Avoid use wherever possible.	Continual reuse of reusable equipment is permitted where adequate decontamination can be assured. ie minimum of 18 minutes autoclaving at 134°C or soaking in 1 – 2M NaOH for 1 hour. For tonometers , protective covers are recommended and should be discarded after use. Non contact air or puff tonometers which totally avoid contact with the cornea should be used. Endoscopes Disinfect according to normal procedures.

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Table 4: continued

Specific recommendations	
Group 1 (high risk)	Group 2 (low risk)
Collection of specimen	
Collect into a secure closing container and enclose in a plastic bag. The container must be labelled clearly with patient details, including a CJD risk alert to laboratory/other medical personnel.	Standard Precautions apply.
Disposal of specimens	
Disposal of all specimens should be by incineration.	Brain and CSF samples should be incinerated. Disposal of other specimens should follow Standard Precautions.
Other materials used in procedures	
Use of reusable articles is not permitted. Swabs, dressings, linen etc used during operations should be incinerated. Needles and other sharps should be placed in appropriate containers for disposal by incineration (in accordance with AS 4031-1992).	All articles coming into contact with blood or CSF should be enclosed in yellow infectious waste bags with international biohazard symbol and the words 'Medical Waste' and disposed of by incineration. Standard Precautions apply for disposal of other waste materials. Needles and other sharps should be placed in appropriate containers for incineration (in accordance with AS 4031-1992).
Anaesthetic equipment	
Discard equipment if contaminated with blood.	Clean according to Standard Precautions.

10.4 Routine hospital/nursing home/domiciliary care

The low potential for transmissibility by non-parenteral routes demonstrates that routine contact with persons in both Groups 1 and 2 does not pose any definite risk. Nevertheless, primary care physicians, nursing home workers, relatives and friends of persons in Group 1 should be aware of the potential infectivity of CSF, blood and serum. Appropriate measures to decontaminate linen soiled with blood or serum should be undertaken.

Table 5: Routine hospital/nursing home/domiciliary care

Specific recommendations	
Group 1 (high risk)	Group 2 (low risk)
Infection control policy	
All major hospitals should have procedures set in place to identify and manage high risk patients. Standard Precautions must also still apply.	Standard Precautions apply in addition to the extra precautions listed below.
Ward accommodation	
Hospital patients need not be accommodated in single rooms.	
Specialist nursing requirements	
isolation and barrier nursing are not required. Staff should, however, be fully informed in the precautions required for the different risk groups.	
Eating utensils	
Cleaning of cutlery, plates, cups etc may be handled as normal. There is no evidence that CJD can be transmitted via saliva.	As for Group 1 patients. No special requirements.
Bath towels and face washers	
Patients should be allocated their own personal face washers and towels. If contaminated with blood, soaking in hypochlorite prior to normal laundering may add an extra margin of safety.	No special requirements. Wash according to normal laundering procedures.
Razors and toothbrushes	
Razors and toothbrushes must not be shared. Dispose of old blades and brushes by incineration.	Razors and toothbrushes should not be shared.
Laundering of normal bed linen and clothes	
Laundering may be as normal except when contaminated with blood or CSF. Linen and clothes soiled with blood or CSF should be incinerated. For this reason it is advisable to use single use absorbent sheet covers wherever practical.	Wash linen and clothes according to normal laundering procedures.
Disposal of contaminated materials	
Ward materials (linen, swabs etc) contaminated with blood, CSF or other bodily wastes should be placed in yellow infectious waste bags with international biohazard symbol and the words 'Medical Waste' for incineration. Needles and other sharp articles should be placed in appropriate biohazard containers for incineration (in accordance with AS 4031-1992).	Ward materials (eg swabs, dressings) contaminated with blood or CSF should be placed in yellow infectious waste bags with international biohazard symbol and the words 'Medical Waste' for incineration. Needles and other sharp articles should be placed in appropriate biohazard containers for incineration (in accordance with AS 4031-1992).

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Table 5: continued

Specific recommendations	
Group 1 (high risk)	Group 2 (low risk)
Collection of specimens	
Blood, tissue or CSF specimens should be collected into sealable containers and labelled clearly with the patient details, including CJD risk status.	Standard Precautions apply.
Cleaning contaminated surfaces – spills	
For surfaces contaminated with blood or body fluids, absorb liquid onto paper towels and place these immediately into incineration container. Soak the area with 1–2 M NaOH or 2.0 – 2.5 per cent sodium hypochlorite and leave for 1 hour. Clean area again with paper towels and incinerate. Also incinerate gloves used for cleaning.	Use caution when cleaning up spills involving blood or CSF. Procedures as described for Group 1 may not be essential, but are preferred. Wear gloves and protective clothing.

10.5 Dentistry

For patients in Group 1 (high-risk), dental procedures in which bleeding occurs should be carried out only by personnel who are familiar with the CJD infection control procedures, and probably at a central referral facility (such as a specialist dental hospital or a dental unit in a major hospital). Instruments contaminated with blood or neurovascular tissue should be disposed of by incineration.

Dentists should take an appropriate medical history of all patients. Persons in Group 2 (low risk) should advise their dentists of their status so that adequate decontamination procedures for reusable equipment can be arranged. The following table provides a guide to the precautions necessary when treating Group 1 and 2 patients.

Table 6: Dentistry

Group 1 (high risk patients)	Group 2 (low risk patients)
Standard Precautions apply in addition to the extra precautions listed below.	
Wherever possible, single use items/equipment should be used.	
During all dental procedures staff should wear masks, protective eye-wear, single use gloves and gowns.	
Procedures involving blood or neurovascular tissue	
<p>Instruments contaminated with blood or neurovascular tissue should all be disposed of by incineration.</p> <p>Dental broaches and burrs, and other items which may have come into contact with blood and neurovascular tissue should also be disposed of by incineration (see Appendix 1).</p> <p>Continual use of reusable equipment is not advised, therefore reusable equipment should be discarded after use on a Group 1 patient.</p> <p>Contaminated handpieces should be disposed of after use.</p>	<p>For procedures involving blood and neurovascular tissue, re-usable instruments should only be used when adequate decontamination procedures are available.</p> <p>Instruments may be decontaminated by:</p> <ul style="list-style-type: none"> - autoclaving at 134°C (30 psi or 203 kPa) for 18 minutes (holding time at temperature) <p>OR - six separate 3 minute cycles (total holding time 18 minutes) at 134°C (30 psi or 203 kPa)</p> <p>OR - soaking in 1–2 M NaOH, minimum 1 hour (NB only suitable for stainless steel instruments).</p> <p>Use single use equipment wherever possible.</p> <p>Contaminated handpieces should be autoclaved as above.</p> <p>Broaches, files etc. cannot be cleaned adequately and therefore must be disposed of after single use (see Appendix 1).</p>
Needles/syringes	
Use single-use needles, syringes and anaesthetic cartridges only.	<p>Single-use needles, syringes and anaesthetic cartridges are recommended.</p> <p>Metal reusable local anaesthetic syringes should be autoclaved as for other stainless steel instruments or soaked in 1–2 M NaOH (see above).</p>

10.6 Laboratory personnel^{14,20a,22,23}, mortuary attendants and funeral industry workers

10.6.1 Laboratory personnel

In the clinical pathology laboratory, specimens from both Group 1 and 2 patients should be treated according to Standard Precautions. However, in the anatomical/surgical pathology laboratory, appropriate containment and decontamination procedures are necessary when handling brain tissue and other surgical specimens from either Group 1 or 2 patients¹⁴. Cut-up/hooking of tissue samples from either infectious category should be performed in a biohazard hood, preferably located in a separate area. Because of the known resistance of CJD infectivity to aldehydes¹⁴ and alcohols, the safest manner in which to handle biopsy material is by fixation of small blocks of tissue followed by immersion in formic acid for one hour²⁰. After washing, these blocks can then be processed routinely for histology.

10.6.2 Post mortem examinations

Guidelines relating to the conduct of post mortem examination in CJD have been reported^{6,7,10}, and should now follow the same protocols established for other infectious diseases such as HIV and tuberculosis. In general, it is recommended that at least one centre in each Australian capital city be designated as a referral centre with expertise in the conduct of such an autopsy. Removal of the brain⁷ is accomplished within the containment of a clear plastic bag to avoid aerosol contamination with the electric bone saw. After immersion fixation of the brain in formalin, blocks can be taken and decontaminated in formic acid as previously described (see 10.6.1). If frozen tissue is to be retained for genetic testing, diagnosis or research, then appropriately labelled containers should be used.

10.6.3 Mortuaries and funeral industry workers

Decontamination of instruments and surfaces in the mortuary should follow the guidelines set out above for Group 1 and 2 patients.

Funeral industry workers should also be aware of these recommendations and take appropriate steps to decontaminate working surfaces and instruments within their mortuaries. Embalming of bodies in Group 1 or 2 should be avoided.

Table 7 :Laboratory

Minimum requirements	
Group 1 (high risk)	Group 2 (low risk)
Standard Precautions	
Standard Precautions should be followed for all procedures performed on Group 1 or 2 patients.	
Autoclaving facilities	
Facilities for autoclaving and chemical decontamination of instruments and surfaces should be available in proximity to the cut-up area and mortuary.	
Protective clothing	
Single use gloves should be worn for all procedures involving contact with body fluids or tissues.	
Gowns (preferably single use), masks and protective eye-wear should be worn, particularly where splashing of blood, tissue and other body fluid may occur.	
Where heavy soiling of non-disposable gowns occurs (by blood, CSF or body tissues), dispose of by incineration.	
Cleaning of contaminated surfaces (eg bench tops, floors)	
Use single use absorbent bench-coat or other bench coverings wherever possible, and dispose of by incineration.	
Where surfaces are contaminated with brain tissue, other neural tissue, CSF or blood, wipe areas with paper towels and incinerate. Drench surface with 1 – 2 M NaOH and leave for one hour. Clean up with paper towels and incinerate.	
Specimen preparation and handling	
Label all specimens with patient details. In anatomical/surgical pathology laboratories, special attention should be given in handling of brain tissue and other surgical specimens from both Group 1 and 2 patients ¹⁴ .	
A separate area with a biohazard hood should be available for cut-up/blocking of tissue samples from Group 1 and 2 patients. Because of the known resistance of infectivity to aldehydes ¹⁴ and alcohols, biopsy material should be fixed in 4 per cent formaldehyde solution (10 per cent formal-saline), followed by immersion in formic acid (> 96 per cent) for one hour. For machine processing, tissues should be re-washed in formalin as formic acid may damage plastic containers. Where tissues are processed by hand, transfer directly from formic acid into ascending alcohol solutions.	
When frozen sections are required, decontamination of the cryostat microtome is required.	
All specimens should be treated as potentially infectious for CJD until proven otherwise.	
Dispose of tissues by incineration.	
NB Do not autoclave formaldehyde solutions.	
Use of cadavers for teaching purposes	
Departments of Anatomy and Pathology should not use cadavers of Group 1 or 2 individuals for teaching purposes.	

11. Organ, tissue and blood donations; therapeutic usage of products of human origin

The following persons should be excluded from the routine donation of organs, tissues and blood:

- persons in risk Groups 1 or 2;
- persons who die in psychiatric hospitals; and
- persons who die with any obscure undiagnosed neurological disorder^{2,41}

It might be argued that tissue donation from the low risk group of persons could proceed if

- 1) the recipient were elderly;
- 2) the recipient were fully informed of the potential risk; and
- 3) the purpose of the transplant procedure were a matter of life or death for the recipient.

Institutional ethics committees should examine this question and reach their own decision.

Persons who are responsible for recruiting organ/tissue donors and for the banking of tissues (eg corneas, heart valves, skin) should be aware of the problem of CJD, and should have exclusionary criteria and procedures in place⁴¹.

In all cases where materials for transplantation, grafting or tissue banking are derived from post mortem material, it is mandatory that the brain of the donor be assessed and cleared by a specialist neuropathologist. Paraffin blocks of brain tissue on such donors should be archived for future reference.

Materials from the above patient groups (Section 11, paragraph 1) should not be used for the preparation of any therapeutic products or laboratory reagents (eg thromboplastin or Kveim test material)^{30,31}. The possible illegal use of contaminated products should also be borne in mind⁵⁰.

12. Genetic testing in familial diseases

Although rare, familial clusters of CJD and GSS have been identified in Australia. Suspected instances of FFI and thalamic dementia are also known. Since most familial diseases carry characteristic mutations in the gene coding for PrP, it is now possible to offer predictive testing, carrier detection and prenatal diagnosis.

Because of the implications of genetic diagnosis, it is recommended that one or more referral centres be organised at which appropriate laboratory testing can be performed, coupled with genetic counselling by persons with expertise and a current knowledge of these diseases.

As indicated above, persons with proven pathogenic mutations or all non-genotyped members of a family in which a clustering of CJD/GSS/FFI occurs must be considered as Group 1 (high risk) individuals and should be advised to take appropriate precautions.

13. Surveillance: The CJD case registry

To assist in the continued surveillance of trends in the incidence and prevalence of CJD, and to identify risk factors associated with the occurrence of CJD, a national CJD Case Registry has been set up and funded by the Commonwealth Department of Human Services and Health. The Registry is based in the Department of Pathology, the University of Melbourne. Since CJD is currently not a notifiable disease, the CJD Case Registry ascertains the occurrence of CJD through a program of periodic contacts with neurologists and anatomical pathologists. It also undertakes surveillance of death certificates and discharge diagnosis from hospitals. Upon notification of a proven or suspected case of CJD, a specialist neurologist and research nurse working for the Registry assess the clinical and pathological information and seek the co-operation of the relatives in completing a questionnaire designed to cover all known or suspected risk factors.

All proven or suspect cases of CJD in Australia should be notified to:

The CJD Case Registry
Department of Pathology
The University of Melbourne
Parkville Vic 3052

Tel: (03) 9344 5868
Fax: (03) 9344 4004

14. Pituitary hormones section

Because of the possible contamination³ with CJD of supplies of human pituitary-derived gonadotrophins (used for treatment of infertility) and growth hormone (used for the treatment of small stature) during the period 1960–1985, the Commonwealth Department of Human Services and Health established a Task Force to trace, inform and counsel recipients of these medications. The Task Force has now become part of the Public Health Division of the Department of Human Services and Health. Persons who believe that they may have been treated with these products may communicate with the Task Force at the following address:

Pituitary Hormones Section
Department of Human Services and Health
GPO Box 9848
Canberra ACT 2601

Tel: 1800 802 306
Fax: (06) 289 6803

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Appendix 1

Other guidelines and standards

In interpreting and implementing the Creutzfeldt-Jakob disease and Other Human Transmissible Spongiform Encephalopathies: Guidelines on Patient Management and Infection Control, readers should be aware of the following guidelines.

AS 4031-1992: Non-reusable containers for the collection of sharp medical items used in health care areas (ISBN 0 7262 7491 7).

AS 4187-1994: Code of practise for cleaning, disinfecting and sterilising reusable medical and surgical instruments and equipment, and maintenance of associated environments in health care facilities (ISBN 0 7262 8849 7).

Draft – NHMRC Infection Control in the Health Care Setting publication, August 1995.

Draft – NHMRC National Guidelines for the management of clinical and related wastes publication, 1995.

Disposal of biological waste

In the absence of an Australian Standard, disposal of biological waste infected with CJD is to be in accordance with best practice or equivalent standards prescribed under the laws, procedures, Codes of Practice or other regulatory provisions in force in the relevant State or Territory that are most consistent with this Guideline.

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The Council comprises nominees of Commonwealth, State and Territory health authorities, professional and scientific colleges and associations, unions, universities, business, consumer groups, welfare organisations, conservation groups and the Aboriginal and Torres Strait Islander Commission.

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