

Guideline Title: CREUTZFELDT-JAKOB DISEASE (CJD) OR OTHER TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHIES (TSE'S)

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GUIDELINES

These guidelines should be read in conjunction with the Infection Control Requirements Policy and other guidelines in the Infection Control Manual available in every ward and clinical department and on the Bayside Health Intranet.

PURPOSE

The purpose of this guideline is to minimise the risk of transmission of Creutzfeldt-Jakob Disease (CJD) and other transmissible spongiform encephalopathies (TSEs).

BACKGROUND

CREUTZFELDT-JAKOB DISEASE (CJD) OR OTHER TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHIES (TSEs)

- The agents that cause Creutzfeldt-Jakob Disease (CJD), new variant CJD (nvCJD), and other transmissible spongiform encephalopathies (TSEs) are prion proteins.
- Prions are a small proteinaceous infectious particle. A protease-sensitive normal host cellular protein (PrP^{sen}) undergoes a structural change to a protease-resistant protein (PrP^{res}); this abnormal protein then accumulates in neural cells, disrupting cell functioning and eventually causing cell death.
- Prions are resistant to a number of standard sterilisation and disinfection procedures.
- There are human and animal TSE's. The HUMAN TSE's are:
 - Creutzfeldt-Jakob Disease (CJD) including classical sporadic; familial; iatrogenic (resulting from medical treatment) and new variant
 - Gerstmann Sträussler Scheinkler syndrome (GSS)
 - Fatal Familial Insomnia (FFI); and
 - Kuru.
- The human TSE's have a pre-clinical phase that lasts for years. This is followed by rapid progressive dementia, loss of memory and intellect, personality changes or progressive unsteadiness and clumsiness. In most cases, death occurs within a few months of onset of symptoms.

- All human TSEs are very rare; the worldwide incidence of CJD is about 1 per million people each year.
- In 1996, a new variant CJD (vCJD) was reported.
- As of December 1, 2003, a total of 153 cases of vCJD had been reported in the world: 143 from the United Kingdom, six from France, and one each from Canada, Ireland, Italy and the United States (Note: the Canadian, Irish, and US cases were reported in persons who resided in the United Kingdom during a key exposure period).
- Almost all the 153 vCJD patients had multiple-year exposures in the United Kingdom between 1980 and 1996 during the occurrence of a large UK outbreak of bovine spongiform encephalopathy (BSE, commonly known as mad cow disease) among cattle.
- It is believed that the persons who have developed vCJD became infected through their consumption of cattle products contaminated with the agent of BSE.

DIAGNOSIS

- Physicians suspect a diagnosis of CJD on the basis of the typical signs and symptoms and progression of the disease.
- A test called the 14-3-3 protein in the CSF is a marker for some prion diseases such as Creutzfeldt-Jakob Disease, when a number of other conditions are excluded.
- A confirmatory diagnosis of CJD requires neuropathologic and/or immunodiagnostic testing of brain tissue obtained either at biopsy or autopsy.
- Brain biopsy is *not* recommended for diagnostic purposes.

IATROGENIC TRANSMISSION

- TSEs are *not* known to spread by contact from person to person, but transmission can occur during invasive medical intervention.
- Iatrogenic transmission of the CJD agent has been reported in over 250 patients worldwide. These cases have been linked to the use of contaminated human growth hormone, dura mater and corneal grafts, or neurosurgical equipment.
- Of the six cases linked to the use of *contaminated equipment*, four were associated with neurosurgical instruments, and two with stereotactic EEG depth electrodes.
- All of these equipment-related cases occurred before the routine implementation of sterilisation procedures currently used in hospitals. No such cases have been reported since 1976.

INFECTIVITY OF HUMAN TISSUES

This information is based largely on studies of experimentally transmitted CJD in non-human primates and other animals.

INFECTIVITY CATEGORY	TISSUES, SECRETIONS, EXCRETIONS	
HIGH INFECTIVITY	Brain Spinal cord Eye (retina and optic nerve)	
LOW INFECTIVITY	Cerebrospinal fluid (CSF) Lung Liver Kidney Spleen/lymph nodes Placenta	
NO DETECTABLE INFECTIVITY	Heart Peripheral nerve Gingival tissue Adrenal gland Prostate Urine Sputum Mucous Milk Sweat Blood*	Skeletal muscle Adipose tissue Intestine Thyroid Testis Faeces Saliva Semen Tears Serous exudate

* Experimental transmission of CJD by blood has been demonstrated in animal models. In the UK, transfusion associated variant CJD has been reported, however the risk of transmitting variant CJD through blood remains uncertain.

PROCEDURES

- **STANDARD PRECAUTIONS** *must* be followed when caring for patients with suspected CJD or other transmissible spongiform encephalopathies (TSEs) (see Standard Precautions Guideline).
- All staff *must* adhere to the following:

A. NOTIFICATION REQUIREMENTS (see Appendix 1)

- All staff *must* notify Infection Control of any patients suspected or confirmed to have Creutzfeldt-Jakob disease (CJD) or other transmissible spongiform encephalopathies (TSEs).

B. PATIENT PLACEMENT

- Patients suspected or confirmed to have Creutzfeldt-Jakob disease (CJD) *do not* require single rooms for infection control purposes.

C. OPERATIVE PROCEDURES (see Appendix 1)

- Prior to booking an elective operative procedure, patients suspected of having CJD or another transmissible spongiform encephalopathy (TSEs) **must** have a neurology consultation.
- When booking the patient for an operative procedure, the Medical Officer **must** notify the Operating Suite Floor Coordinator that the patient is suspected of having CJD or another transmissible spongiform encephalopathy (TSEs).
- Before the patient is transferred the ward/unit in which the patient is being cared for **must** notify the Operating Suite Floor Coordinator that the patient is suspected of having CJD or another transmissible spongiform encephalopathy (TSEs).
- The operating theatre staff **must** follow policies and guidelines outlined in the Operating Suite Policy Manual – **Safe Practice and the Environment 8: Management of Patients with Potential/Confirmed Transmissible Spongiform Encephalopathies.**
- Pre-planning relating to the following is required:
 - Procedure scheduling
 - Staff notification (eg anaesthetic, ancillary, laboratory, OSSA and CSSD)
 - Staff personnel protective equipment (PPE)
 - Supplies, equipment and instruments to be used
 - Anaesthetic equipment
 - Instrument handling
 - Specimen handling
 - Instrument cleaning, decontamination and sterilisation
 - Instrument and specimen transfer between departments
 - Instrument quarantining
 - Monitoring and maintenance processes of quarantined instruments
 - Environment cleaning
 - Waste and sharps disposal
- Surgical procedures performed in the Operating Theatre **must**:
 - Involve minimum required number of staff
 - Use single use equipment as follows:
 - Liquid repellent operating theatre gowns over a plastic aprons
 - Gloves
 - Mask
 - Protective eyewear
 - Disposable linen/drapes
 - Single use disposable instruments where possible.

D. INVASIVE PROCEDURES

- Invasive procedures, which are normally carried out at the bedside or in procedural areas/departments (eg lumbar puncture, surface electrodes where abrasion of the skin surface occurs) **must** be well planned and cover the above issues.
- The ward/unit **must** notify the accepting area that the patient is suspected of having CJD or another form of transmissible spongiform encephalopathies (TSEs).

E. HANDLING/REPROCESSING AND QUARANTINING INSTRUMENTS (see Appendix 1)

The safest method for ensuring that there is no risk of residual infectivity on contaminated instruments and other materials is to discard and destroy them by incineration.

CONFIRMED CASES

- At the completion of the procedures all **disposable instruments and materials** are to be discarded.
- At the completion of the procedures all **reusable instruments** and materials **must** be **discarded and destroyed by incineration**.

SUSPECTED CASES

- At the completion of the procedures all **disposable instruments and materials** are to be discarded.
- At the completion of the procedures all **reusable instruments** and materials **must** be **quarantined** until the final diagnosis has been established.
- The final diagnosis will be determined by the Infectious Diseases Unit in liaison with specific Units/Departments Heads.
- Infection control maintains a register of all patients who have undergone a surgical or invasive procedure in which the instruments used in the procedure were **quarantined**.
- Quarantined items are **not** to be removed from quarantine **without written authorisation from Infection Control**.

F. INEFFECTIVE OR SUB-OPTIMAL INSTRUMENT REPROCESSING METHODS

- Some commonly used chemicals and processes that **cannot** be depended upon for decontamination as they have been shown to be **ineffective** or only **partially effective** in destroying TSE, are listed in the following table.

CHEMICAL DISINFECTANTS	GASEOUS DISINFECTANTS	PHYSICAL DISINFECTANTS
<p><u>Ineffective</u></p> <p>Alcohol Ammonia Formalin Hydrochloric acid Hydrogen peroxide Peracetic acid Phenolics</p> <p><u>Variable or partially effective</u></p> <p>Chlorine dioxide Gluteraldehyde Iodophores</p>	<p><u>Ineffective</u></p> <p>Ethylene oxide (ETHO) Formaldehyde</p>	<p><u>Ineffective</u></p> <p>Boiling Dry heat (<300°) Ionising, UV or microwave radiation</p> <p><u>Variable or partially effective</u></p> <p>Autoclaving at 121°C for 15 minutes</p>

G. DECONTAMINATION METHODS FOR QUARANTINED INSTRUMENTS

- If sterilisation methods are employed prior to quarantining, non-disposable instruments/equipment, *two or more different methods of inactivation are to be utilised*.
- All items *must* be dismantled and mechanically cleaned prior to decontamination and sterilising.

METHODS

<p>AUTOCCLAVING</p> <p>Prevacuum autoclave. 134°C (pressure 203 kPa at 134°C). Holding time at temperature should be at least 18 minutes single cycle or six separate 3-minute cycles.</p>
<p>EXPOSURE TO 1-2 M SODIUM HYDROXIDE</p> <p>1M NaOH equals 40 g NaOH per one litre of water. Expose instruments to this solution for at least 1 hour at room temperature. Completely submerge the instruments. Physically scrub the instruments to remove excess tissue/blood while in the NaOH. Ensure all items used for cleaning are adequately decontaminated.</p>
<p>SOAKING IN SODIUM HYPOCHLORITE</p> <p>Freshly prepared Hypochlorite solutions should contain 2.0 to 2.5 per cent sodium hypochlorite. Soak instruments for at least 1 hour.</p>

H. DECONTAMINATION OF WORK SURFACES

1. The work area should be selected for easy containment of tissue/secretions and excretions.
2. Use disposable cover sheets wherever possible.
3. All waste liquids and solids *must* be captured and treated as infectious waste.

4. Surfaces contaminated with tissue/secretions and excretions from patients with suspected or confirmed TSE, can be disinfected by flooding the area, for 1 hour with Sodium Hydroxide (2N NaOH) or sodium hypochlorite (undiluted) followed by rinsing with water.
5. Where surfaces cannot tolerate NaOH or sodium hypochlorite, thorough cleaning will remove most infectivity by dilution and some additional benefit *may* be derived from the use of one or other of the partially effective methods listed above.
6. All individuals involved in disinfection and decontamination procedures should be familiar with basic procedures and wear personal protective equipment (PPE).

I. WASTE DISPOSAL

- Waste *must* be disposed of in accordance with the Waste Management Policy available on the intranet.

J. LABORATORY OR PATHOLOGY SPECIMENS

- All specimens from known or suspected cases of TSE *must* be collected in plastic screw top specimen containers.
 - Place the specimen in a second plastic screw top container.
 - Label with a biological hazard sticker.
 - Insert into the pathology request form and seal.
 - Transported to Alfred Anatomical Pathology Department.
- Transport of the specimen off site *must* be arranged through the Anatomical Pathology Department.
- Dr McLean, Consultant Pathologist or the Head of the Anatomical Pathology Department at The Alfred *must* be notified well before the procedure commences.

K. PRECAUTIONS FOR HANDLING THE DECEASED

- On the death of a patient with suspected or confirmed case of CJD or other TSE the removal of the body from the ward should be carried out using routine infection control measures (see Standard Precautions Guideline).

L. POST MORTEM EXAMINATION

- Requests and enquiries regarding post mortem examination are to be made with Dr McLean, Consultant Pathologist at The Alfred or the Head of the Anatomical Pathology Department.

M. CJD EXPOSURES

- All needlestick, sharps and splash exposures *must* be reported immediately (see Needlestick/Sharp Injury or Blood/Body Fluid Exposure Guideline).

APPENDICES

Appendix 1: Handling/Reprocessing and Quarantining Instruments Used For Surgical and Invasive Procedures

RELATED DOCUMENTATION

Standard Precautions Guideline	Intranet
Needlestick/Sharp Injury or Blood/Body Fluid Exposure Guideline	Intranet
Waste Management Policy	Intranet

REFERENCES

1. Infection Control Guidelines for the Prevention of Transmission of Infectious Diseases in the Healthcare Setting. 2004. Australian Government, Department of Health and Ageing.
2. WHO Infection Control Guidelines for Transmissible Spongiform Encephalopathies. Report of a WHO consultation, Geneva, Switzerland, 23-26 March 1999.
http://www.who.int/csr/resources/publications/bse/WHO_CDS_CSR_APH_2000_3/en/
3. Fact Sheet - New Variant Creutzfeldt-Jakob Disease. CDC. January 2004
http://www.cdc.gov/ncidod/diseases/cjd/cjd_fact_sheet.htm

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HANDLING/REPROCESSING AND QUARANTINING INSTRUMENTS USED FOR SURGICAL AND INVASIVE PROCEDURES

