Washington State Department of Health
DOH 420-162

HUMAN PRION DISEASE INFECTION CONTROL KEY POINTS

Background

Prion diseases, also referred to as transmissible spongiform encephalopathies (TSE), are a rare group of progressive neurodegenerative disorders that can occur in humans and animals. The presence of an abnormally folded protein, called scrapie prion protein (PrP^{Sc}) is required to definitively diagnose prion disease.

Creutzfeldt-Jakob disease (CJD) is the most common human prion disease. It is a rare, fatal disease commonly characterized by rapidly progressing dementia, poor balance, visual changes and/or muscle jerks. Sporadic CJD (sCJD) has no known cause and accounts for about 85% of all CJD cases. Familial CJD (fCJD) results from an inherited mutation and accounts for 10–15% of cases. Very rarely, CJD is acquired. A new variant CJD (vCJD) that was recognized in the United Kingdom in 1996 has now been associated with eating beef products from cows affected with bovine spongiform encephalopathy (BSE, "mad cow disease"). As May 2016, 227 vCJD have been reported globally. Four cases of vCJD have been diagnosed in the United States, but were acquired elsewhere.

To date, no cases of variant CJD are thought to have been acquired in Washington or the United States.

Iatrogenic CJD (iCJD) is an uncommon acquired form of prion disease. Iatrogenic transmission of the CJD agent has been reported in over 450 patients worldwide. These cases have been linked to the use of contaminated human growth hormone (hGH) extracted from cadavers prior to 1977 (29 cases in the U.S.), dura mater (4 cases in the U.S.) and corneal grafts (1 case in the U.S.), or neurosurgical equipment (no cases in the U.S.)[4]. 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 known equipment-related cases occurred before the routine implementation of sterilization procedures currently used in health care facilities. No such cases have been reported since 1976. No iatrogenic CJD cases associated with exposure to the CJD agent from surfaces such as floors, walls, or countertops have been identified. The only case of iCJD that has been reported in Washington State (in 2013) was associated with hGH administration during childhood.

For information regarding human prion disease epidemiology in Washington State, please see Annual and Decennial Prion Disease Surveillance data.

Human prion disease is a notifiable condition in Washington State (WAC 246-101-101) and all suspected and confirmed cases should be reported to your Local Health Jurisdiction.

Infection Control Considerations

- There is no evidence that normal social or routine contact with a CJD patient represents a risk for healthcare workers, household members, or others. Isolation of patients with CJD is not necessary. Standard precautions should be used for all patients with known or suspected CJD.
- Body secretions and body fluids are all low risk for the presence of the CJD agent. It is therefore likely that the majority of samples taken or procedures performed will be low risk for transmission (See Table 1). Contact with small amounts of blood (including injury by inoculation) is also considered low risk, although transfusion of large volumes of blood has rarely been associated with transmission (vCJD only).

- If a patient with suspected or confirmed prion disease requires an invasive procedure, healthcare providers should contact the facility's infection control division to assure that recommended infection control measures are implemented.
- Although no equipment-related iCJD cases have been reported since implementation of the currently-used sterilization procedures, this may reflect the inefficiency of transmission unless neural tissue is involved. The abnormally folded prion proteins are still considered to be highly resistant to standard decontamination and sterilization methods.
- No single method is considered to be 100% effective against prions but instruments in contact with certain high risk tissues (i.e. brain, spinal cord, and eye) can be decontaminated by a combination of specific chemical and autoclaving methods before subjecting them to routine sterilization.
- It is important that only trained staff, who are aware of the hazards, carry out invasive procedures that may lead to contact with high risk tissues.
- Patients with known or suspected prion disease should not serve as donors for organs, tissues, blood components, or sources of tissue (e.g. dura mater and hormones).

Table 1. Infectivity in Organs, Tissue, and Body Fluids of Humans with Prion diseases (CJD)

Infectivity category	Tissues, secretions, and excretions			
High infectivity	Brain	Pituitary gland	Cranial nerves	
	Spinal cord	Posterior eye	Cranial ganglia	
Low infectivity	Cerebrospinal Fluid (CSF)		Lung	Liver
	Lymph nodes/spleen		Placenta	Kidney
	Olfactory epithelium			
No detectable infectivity	Adipose tissue	Heart muscle	Peripheral nerves	Feces
	Skeletal muscle	Blood /serum	Nasal mucous	Prostate
	Adrenal gland	Semen	Testis	Saliva
	Serous exudate	Intestine	Sweat	Milk
	Gingival tissue	Tears	Thyroid gland	Urine
	Bone marrow	Sputum	Vaginal secretions	

Recommendations for disinfection and sterilization of prion-contaminated medical devices

- Instrument reprocessing should be planned well in advance of procedures done on patients with known or suspected CJD.
- Neurosurgical instruments used to treat patients whose diagnosis is unclear, particularly for brain biopsy, should be regarded as potentially contaminated with the CJD agent. Such instruments should be quarantined until a non-prion disease diagnosis is identified or should be regarded as contaminated and sterilized using the recommended CJD de-contamination protocols.
- Instruments should be kept wet or damp after use (using water or a prionicidal detergent) and decontaminated as soon as possible.
- Instruments should first be manually cleaned, then decontaminated by a combination of the recommended chemical and autoclaving methods, and finally subjected to cleaning in a washer cycle and routine sterilization. (Before instruments are immersed in any chemical agent, the instrument manufacturer should be consulted about the instrument's tolerance of exposure to the agent.)
- Prion-contaminated medical devices that are impossible to clean or fully expose to steam and other sterilants should be discarded.

- Always discard single-use devices.
- Surfaces should be covered with plastic-backed paper, and if the paper has been contaminated with high-risk tissues, it should be properly discarded.
- Surfaces contaminated with tissues from CJD patients should be cleaned and then decontaminated with a 1:10 dilution of hypochlorite solutions, ideally for a contact time of at least 15 minutes.
- All disposable instruments, materials, and wastes that come in contact with high and low infectivity tissues of suspected or confirmed TSE patients should be disposed of by incineration or as hazardous waste.
- For detailed information on managing suspected or confirmed cases including specific instructions on chemical and autoclaving methods available to sterilize prion-contaminated equipment, please refer to Washington State Department of Health Human Prion Disease guideline (Section 5.B).

Other recommendations

- Ideally, institutions should have a tracking system in place that facilitates recall of critical or semi-critical devices used on high-risk tissue and high-risk patients. This tracking system should permit identification of each patient on which the devices were used including the date of use, procedure performed, and surgeon's name.
- Any decision to notify patients on whom such instruments were used should be based on an analysis of the risk of CJD transmission.
- If not already in place, CJD infection prevention guidelines tailored to your institution should be developed and periodically updated as new information becomes available.

References/Resources

1. World Health Organization (WHO) Infection Control Guidelines for Transmissible Spongiform Encephalopathies for suspected or confirmed human prion disease: http://www.who.int/csr/resources/publications/bse/whocdscsraph2003.pdf
2. A summary of the management of exposed patients to inadequately sterilized neurosurgical instruments contaminated is available at http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4748700/pdf/nihms-723714.pdf
3. Additional information about infection control measures related to CJD is available from the Centers for Disease Control and Prevention at http://www.cdc.gov/prions/cjd/infection-control.html
4. Brown P, Brandel J-P, Sato T, et al. Iatrogenic Creutzfeldt-Jakob Disease, Final Assessment. Emerging Infectious Diseases. 2012;18(6):901-907. doi:10.3201/eid1806.120116.

Questions?

If you have any questions regarding these Key Points about infection control procedures for prion disease, please contact the Washington State Department of Health prion disease epidemiologist at: (206) 418-5500.