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Policy Area Infection Prevention
Applicability UNC Medical Center

Creutzfeldt-Jakob Disease (CJD)

I. Description

Describes the recommendations to prevent the transmission of sporadic Creutzfeldt-Jakob Disease (CJD) within the healthcare setting

II. Rationale

CJD is transmitted by a proteinaceous infectious agent or prion. The CJD prion is hardy and resistant to heat, formaldehyde, glutaraldehyde, ionizing radiation, freezing, drying, and organic detergents. Because it resists routine cleaning, disinfection, and sterilization procedures used in hospitals, diligent adherence to the following recommended practices, specifically for surgical instrument handling, is essential. CJD has been shown to be transmitted between humans by only certain tissues that include infected brain, ocular tissue, and pituitary tissue/hormones. "For a surgical instrument to act as a vehicle of prion transmission, it must come into contact with infective tissue (e.g., brain) during surgery of the infected patient, it must retain the infectivity of an adhered matter after being decontaminated and sterilized, and it must have contact with the receptive tissue in the recipient."³

III. Policy

A. Patients at High-Risk of Having CJD

1. Patients with known or suspected prion diseases (e.g., CJD, Gertsmann-Straussler Sheinker [GSS] disease, fatal familial insomnia [FFI syndrome]).
2. Rapidly progressive dementia consistent with possible prion diseases.
3. Patient is undergoing brain biopsy when a specific lesion has not been demonstrated (e.g., via MRI, CT).

4. Familial history of CJD, GSS, or FFI.
5. Patients known to carry a mutation in the PrP gene involved in familial Transmissible Spongiform Encephalopathies (TSEs).
6. EEG showing periodic, sharp wave complexes suggestive of CJD with a clinical history consistent with CJD.

B. Infection Prevention Measures

1. Communication

a. Attending Physician:

- i. Notify Infection Prevention and other involved departments (e.g., Perioperative Service, Neurophysiology, Pathology, Central Processing Department, Microbiology) when a patient with known or suspected prion disease or at high risk for CJD is scheduled for any invasive procedure in which there may be exposure of instruments to high-risk tissues.
- ii. In the event of patient death, the physician will ensure that the morgue and funeral home are notified that the patient has known or suspected CJD.

b. Infection Prevention:

- i. After notification of a patient with known or suspected CJD (by lab or Attending): investigate if the patient had or is scheduled to have a brain biopsy or other surgical procedure that may involve contact with high-risk tissues. If surgery occurred or is scheduled, notify the following:
 - **OR:** Advise personnel on appropriate transport and sterilization of all reusable instruments. Advise personnel preparing specimens for transport to Anatomic Pathology that all specimen containers must be properly labeled and tightly closed. Special procedures for fixing tissues possibly infectious with CJD are required.
 - **CPD:** Advise personnel on appropriate transport and sterilization of all reusable instruments. If a brain biopsy or other procedure involving high-risk tissues has been done prior to diagnosis, CPD will be instructed to track all instruments for recall.
 - **Anatomic Pathology:** Notify the Histology Supervisor at

984-974-1478 of specimen coming to the lab including the patient's name and differential diagnosis. A formalin-formic acid procedure is required for inactivating virus infectivity in tissue samples from patients with CJD.

- **Neurophysiology:** EEG leads contaminated with high-risk tissues must be sent to Central Processing Department and are to be sterilized as described below (3Bi). Alternatively, they may be discarded in a needle disposal container. EEG leads that have had only skin contact may be reprocessed in the conventional manner.
- **Clinical Microbiology Laboratory:** 984-974-1805

2. General Precautions for Patients with Known or Suspected CJD

a. Additional Precautions

- i. Place Epic flag in Infection Status for patients with known or suspected CJD. This flag may be removed if CJD is ruled out.
- ii. Label high-risk tissues (brain, spinal cord, posterior eyes, pituitary tissue) "BIOHAZARD" and "suspected CJD" prior to transport.
- iii. Autopsies are not performed on patients with known or suspected CJD at UNCMC. The UNC Hospitals' morgue should follow Standard Precautions. Lab-Autopsy CJD/TSE Policy
- iv. Patients with known or suspected prion disease should not serve as donors for organs, tissues, blood components, or biologic products (e.g., interferon). Recipients of pituitary hormones or dura mater transplants should not be donors. Transfusion/ Transplant services should include this factor in pre-donation surveys.

b. Standard Precautions

- i. Follow Standard Precautions utilizing appropriate personal protective equipment (e.g., handle all tissue samples using gloves).
- ii. No special precautions are required for handling laundry, food trays, or utensils. No special precautions are required for disposal of body fluids. Such fluids may be disposed of via a sanitary sewer in accordance with applicable regulations.
- iii. A private room for the patient is not necessary for infection prevention purposes.

- iv. Discard disposable instruments in the appropriate receptacle.
 - v. Manage regulated medical waste (i.e., bulk blood, pathological materials, biologic materials, and sharps) according to the Infection Prevention policy: Guidelines for Disposal of Regulated Medical Waste.
3. Decontamination of CJD Contaminated Medical/Surgical Devices: Decontamination methods are based upon the type of contamination (i.e., high-risk tissues, low-risk tissues/fluids, no risk tissues/fluids), and the potential risk of infection involved in the use of the items.

a. Categories of Risk of Patient-Care Items

- i. Critical Items: Items that enter sterile tissue or contact the vascular system, have a high risk of transmitting infection if contaminated.
- ii. Semi-critical Items: Items that come into contact with mucous membranes or skin that is not intact. (e.g., most endoscopes, respiratory therapy equipment).
- iii. Non-critical Items: Items that touch only intact skin. (e.g., blood pressure cuffs).

b. Decontamination Procedures for Surgical Instruments

- i. High-Risk Tissue (brain, spinal cord, posterior eyes, pituitary tissue) and Critical items from high-risk patients:
 - Discard devices that are difficult to clean.
 - Place instruments in a puncture and leak proof container for transport to Central Processing Department (CPD).
 - Keep instruments moist (either by use of a transport gel/foam, immersion in water, or by use of a wet cloth draped over the instruments) after use and during storage/transport prior to decontamination in CPD. This will retard adherence of material to the medical device.
 - Label the items as "BIOHAZARD" and "CJD" so the CPD employee receiving them will know they require special processing.
 - Inform CPD that they will be receiving instruments used with a patient known or suspected to have CJD prior to transport.

- If a brain biopsy or other procedure involving high-risk tissue was performed prior to diagnosis, CPD will track all instruments for recall.
- The CJD instruments should be decontaminated and reprocessed as soon as possible using the following 3 steps:
 - i. Place instruments in an autoclave at 134°C for greater than or equal to 18 minutes in a pre-vacuum sterilizer.
 - ii. After autoclaving, place the CJD instruments only in the decontamination washer/disinfector.

NOTE: The washer/disinfector uses fresh water for all cycles, therefore at completion it is ready for use with other instruments.
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- iii. After removal from washer/disinfector, assemble/package instruments and autoclave at 134°C for 18 minutes in a pre-vacuum sterilizer.

NOTE: No other instruments/devices may be in the washer basket or sterilizer with CJD-exposed instruments.

- Do not use immediate-use steam sterilization for reprocessing.
 - Discard items requiring low-temperature sterilization (e.g., ethylene oxide, hydrogen peroxide gas plasma).
 - Recall and appropriately reprocess any items contaminated with high-risk tissues that were not processed according to these recommendations (e.g., medical devices used for brain biopsy prior to diagnosis).
- ii. Low/No Risk Tissue (refer to Attachment 1: Comparative Frequency of Infectivity in Organs/Tissue/Body Fluids of Humans with CJD and other TSE) and Critical/Semi-critical Medical Device: These devices can be cleaned and disinfected or sterilized using conventional protocols or heat or chemical sterilization, or high-

level disinfection as appropriate.

4. Cleaning and Disinfection of Contaminated Environmental Surfaces

a. High-risk tissue (brain, spinal cord, posterior eyes, pituitary tissue) Contamination

- Since CJD infectivity persists for long periods on surfaces, it is important to use disposable cover sheets whenever possible to avoid environmental contamination. Mechanically clean and disinfect equipment and surfaces that are subject to potential contamination to prevent environmental build-ups. Surface contaminated by CJD can be disinfected by flooding, for one hour, undiluted sodium hypochlorite (bleach), followed by rinsing with water.

b. Low-risk or no risk tissue or fluid contamination (refer to Attachment 1: Comparative Frequency of Infectivity in Organs/Tissue/Body Fluids of Humans with CJD and other TSE).

- Use standard disinfection procedures (i.e., any EPA-registered disinfectant or a bleach wipe).

5. Intact Skin, Percutaneous or Mucous Membrane Exposure to High-Risk Tissues (brain, spinal cord, posterior eyes, pituitary tissue)

- Intact skin: Wash with detergent and abundant 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.
- Percutaneous (laceration): gently encourage bleeding, wash (avoid scrubbing) with warm soapy water, rinse, dry and cover with a waterproof dressing.
- Mucous Membranes (splashes into the eye or mouth): irrigate with either saline (eye) or tap water (mouth).
- After cleansing the exposed area, the injured health care personnel (HCP) should contact the appropriate occupational health service (Hospital Employees-Occupational Health Service at 984-974-4489 or Needlestick Hotline at 984-974-4480 after hours; University Employees contact University OHS at 984-974-9119 or Healthlink at 984-974-6302 after hours). For large wounds or deep laceration after hours, HCP should be seen in the ED as soon as possible.

6. Contact Infection Prevention at 984-974-7500 (the IP on-call may be reached 24/7 by calling the Hospital Operator or by paging through the hospital directory) for any questions or problems that arise while caring for a patient with suspected or known

IV. References

¹Kavanagh, B. (2014). Creutzfeldt-Jakob disease and other prion diseases. Arlington, VA: Association for Professionals in Infection Control and Epidemiology.

²Rutala, WA, DJ Weber. 2001. Creutzfeldt-Jakob disease: Recommendations for disinfection and Sterilization Healthcare Epidemiology 32: 1348-1356

³Rutala, WA, DJ Weber. 2010. Guideline for disinfection and sterilization of prion-contaminated medical instruments. Infection Control Hospital Epidemiology. 31:107-117.

⁴World Health Organization (1999, March 26). WHO Infection Control Guidelines for Transmissible Spongiform Encephalopathies. Retrieved April 12, 2018, from <http://www.who.int/csr/resources/publications/bse/whocdscsraph2003.pdf>

V. Related Policies

[Infection Prevention Policy: Guidelines for Disposal of Regulated Medical Waste](#)

Attachments

[01: Comparative Frequency of Infectivity in Organs/Tissue/Body Fluids of Humans with CJD and other TSE](#)

Approval Signatures

Step Description	Approver	Date
Policy Stat Administrator	Kimberly Novak-Jones: Nurse Educator	05/2022
	Thomas Ivester: CMO/VP Medical Affairs	05/2022
	Emily Vavalle: Dir Epidemiology	03/2022
	Sherie Goldbach: Project Coordinator	03/2022