SETON HEALTHCARE FAMILY POLICY
Creutzfeldt – Jakob Disease (CJD) and Other Prion Associated Diseases (PAD) Recommendations

☐Administrative ☒Clinical Care ☐Departmental
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Purpose

To provide guidance regarding infection prevention and control practices for healthcare workers involved in the care of patients with suspected or confirmed Creutzfeldt-Jakob Disease (CJD) or other Prion Associated Diseases (PAD) and to prevent transmission to another patient or healthcare worker.

Definitions

A. Cutaneous – Pertaining to the skin
B. Prion – A proteinaceous infectious agent
C. Prion Associated Diseases (PAD) – Fatal illnesses characterized by a rapidly progressive dementia, myoclonus, psychiatric changes, often EEG changes specific to CJD and spongiform neuropathologic changes. Prion associated diseases include: Creutzfeldt-Jakob Disease (CJD), Fatal Familial Insomnia (FFI), Gerstman-Straussler-Scheinker syndrome (GSS), Mad Cow Disease (vCJD).
D. Transcutaneous – Through the skin
E. 14-3-3 – Protein useful in diagnosing CJD. This protein has been reported in 95% of patients with CJD although elevations of this protein also can be seen in patients with other diseases, resulting in a degree of false positivity.

Key Points

A. **Contact Infection Prevention through the hospital operator when a suspected or confirmed CJD/PAD patient requires lab testing, an invasive procedure or surgery.**
B. If a procedure is scheduled to rule out CJD/PAD, infection prevention procedures must be implemented regardless of level of suspicion.
C. Specimens collected to rule out CJD/PAD must be processed and packaged per the National Prion Disease instructions. http://www.cjdsurveillance.com/ (see Resources for Physicians and Referral Labs)
D. All disposable instruments, materials, and waste that come in contact with high risk tissues (brain, spinal cord, and eyes) and low risk tissues (cerebrospinal fluid, kidneys, liver, lungs, lymph nodes, spleen, and placenta) of suspected or confirmed CJD/PAD patients should be disposed of by incineration:
    1. Incinerate all disposable items:
        a. Call SPD or Housekeeping for 2 biohazard bins lined with yellow bag.
        b. Yellow liners/bags are an indication for incineration.

General Information

A. CJD/PAD remains a concern for the healthcare community because of their inherent resistance to traditional disinfection/sterilization methods and devastating clinical outcomes.
B. Transmission of CJD has not been associated with environmental contamination or person-to-person skin contact. Although isolated episodes of CJD have occurred in healthcare workers, the incidence of CJD in this group does not exceed what would be expected by chance alone.

Risk Evaluation

A. Risk is dependent upon three considerations:
    1. The probability that an individual has or will develop CJD
    2. The level of infectivity in tissues of these individuals (**Table 1**)

Table 1: Estimation of Level of Infectivity of Tissues, Secretions and Excretions

Infectivity Risk	Tissues, Secretions and Excretions
High	Brain (including dura mater), Eye, Pituitary tissue, Spinal cord
Low	Cerebrospinal fluid (CSF), Kidney, Liver, Lung, Lymph nodes/spleen, Olfactory epithelium, Placenta
No Detectable Infectivity	Adipose tissue, Adrenal gland, Blood/Leukocytes/Serum, Bone marrow, Feces, Gingival tissue, Heart muscle, Intestine, Milk, Nasal mucous, Peripheral nerve, Prostate, Saliva, Semen, Serous exudates, Skeletal muscle, Sputum, Sweat, Tears, Testis, Thyroid gland, Urine, Vaginal secretions

    3. The nature or route of the exposure to these tissues (**Table 2**)

Table 2: Route of Exposure and Risk

Route of Exposure*	Tissue Infectivity	Risk
Cutaneous - Intact skin Mucous membranes (except those of the eye)	All tissues, secretions and excretions	Negligible risk
Transcutaneous – Non-intact skin Mucous membranes	Low or high infectivity tissues	Unknown – consider a potential risk

Splashes to the eye Inoculations via needle or scalpel or other surgical instruments		
CNS – Inoculation of the eye or CNS	Any infectious material	Serious Risk

*****Healthcare workers with transcutaneous or mucous membrane exposure will be managed as described in the Bloodborne Pathogen - Exposure Control Plan

B. Use risk considerations outlined above to guide decisions about appropriate personal protective equipment (PPE).

Communication

A. When a patient is suspected or known to have CJD/PAD, contact Infection Prevention prior to diagnostic procedure.
B. Infection Prevention is responsible for coordinating communication with lab, surgery and other procedural areas. Infection Prevention shall notify site leaders of confirmed CJD/PAD results.
C. **Infection Prevention is responsible for approving the final release of any quarantined instruments**.
D. All low risk (see Table 1) lab specimens submitted for prion disease testing must be labeled as "CJD or PAD" at the time of collection and hand delivered to the laboratory.
E. Laboratory, OR and Imaging personnel shall have documented annual CJD/PAD education.

Precautions

A. Follow **Standard Precautions** for the care of all suspected or known CJD/PAD. Contamination by body fluids (categorized as no detectable infectivity tissues) poses no greater hazard than for any other patient. Special precautions are NOT required for feeding utensils, feeding tubes, suction tubes, or bed linens.
B. Follow special precautions for diagnostic and surgical procedures outlined below.

Diagnostic Procedures

A. Most diagnostic procedures will not require special precautions, as most tissues with which the instruments come in contact contain no detectable infectivity (see Table 1).

B. **Diagnostic Procedures requiring special precautions:**

1. Lumbar Puncture (Low Infectivity Risk)

a. The National Prion laboratory requests that urine samples accompany CSF submissions.
b. Notify Pathology to deliver National Prion Disease Pathology Surveillance Center paperwork to the department performing the procedure.
c. The department performing the procedure will be responsible for completing National Prion Disease Pathology Surveillance Center paperwork and labeling specimen CJD or PAD. The specimen shall be hand delivered to the laboratory in a biohazard bag.
d. Lab personnel will be responsible for the packaging & shipping of low infectivity risk specimens (see Table 1). See Resources for Physicians and Referral Labs http://case.edu/med/pathology/centers/npdpsc/
e. Equipment & Supplies
    i. Use disposable single-use equipment/supplies.
    ii. Call SPD/Housekeeping for 2 biohazard bins and 2 yellow bags. If biohazard bins are received with red biohazard bags (liners), remove the red biohazard bags and replace with yellow bags. Yellow bags are an indication for disposal through incineration. Only yellow bags may be used for CJD/PAD waste in order to ensure incineration. Red Biohazard bags must never be used.
    iii. Sharps container
    iv. Drape: Cover work surfaces with a disposable impermeable barrier which can then be removed and disposed of in the yellow bags.
    v. Gowns: Wear long-sleeved fluid repellant disposable gowns if performing or assisting with procedure.
    vi. Face shields and Masks: Wear masks with face shields or mask and eye protection goggles if performing or assisting with procedure.
    vii. Gloves: Wear double sterile gloves if performing the procedure.
f. Post Procedure Cleaning
    i. Place used sharps in sharp container & seal.
    ii. Use 1st biohazard bin lined with yellow bag to dispose of sealed sharp container & disposable equipment/supplies.
    iii. Use 2nd biohazard bin lined with yellow bag to dispose of all disposable barriers, patient gown, linen or PPE that came in contact with CSF. Items that did not come in contact with CSF may be placed in trash or linen receptacle as usual.
    iv. Place closed biohazard bin lined with yellow bag in soiled utility room.

v. Follow **Appendix B** for any CSF environmental contamination. If no CSF contamination has occurred, standard cleaning is appropriate.

Dental Procedures

A. Major Dental Procedures
   1. General infection prevention practices recommended by the National Dental Association are sufficient when treating prion infected patients during procedures not involving neurovascular tissue.
   2. CDC "Recommendations from the Guidelines for Infection Control in Dental Health-Care Settings" can be referenced via the following links: https://www.cdc.gov/oralhealth/infectioncontrol/pdf/safe-care2.pdf https://www.cdc.gov/oralhealth/infectioncontrol/pdf/recommendations-excerpt.pdf

Surgical Procedures

A. **Surgeons**
   1. **Surgeons must document suspected or confirmed CJD/PAD cases**.
   2. Scheduling coordinator will contact the surgeon for clarification of unclear bookings. For example, if a Burr hole procedure is booked for "Brain Biopsy: other", then it must clarified that CJD is not in the differential.
   3. Scheduling coordinator notifies Infection Prevention.
   4. The case should be scheduled to allow adequate time for case preparation and decontamination of the environment.

B. **Surgical Services**
   1. The Surgery Department Leader will collaborate with the OR team and Infection Prevention to plan for the procedure regarding instrument handling, storage, cleaning and decontamination or disposal.
   2. Request Lab to deliver National Prion Disease Pathology Surveillance Center paperwork and packaging/shipping instructions and materials to OR.
      a. OR Leader designee will be responsible for completing Prion Center paperwork and the packaging/shipping of the biopsy specimen.
      b. Document in the intraoperative record the packaging actions and transport of specimen to Lab.

C. **General Anesthesia**
   1. Patients with known or suspected vCJD have infectivity detectable in lymphoid tissue such as tonsils; therefore use a disposable laryngoscope or quarantine the laryngoscope
   2. Infection Prevention is responsible for the final release of any quarantined laryngoscopes/instruments.

D. **Handling of Surgical Instruments**
   1. Single-use surgical instruments contacting high infectivity tissues (see Table 1) are strongly recommended and each facility shall maintain a single-use tissue biopsy tray.
   2. See the "WHO Infection Control Guidelines for Transmissible Spongiform Encephalopathies" document for further decontamination recommendations for vital pieces of equipment. http://www.who.int/csr/resources/publications/bse/whocdscsraph2003.pdf?ua=1
   3. Instruments must be quarantined until test results are received from the National Prion Disease Pathology Surveillance Center (see Appendix A). **If single-use instruments are not available and CJD/PAD is confirmed, quarantined instruments shall be destroyed.**
   4. Infection Prevention is responsible for the final release of any quarantined instruments.
   5. Instruments or medical devices that have been contaminated with no-detectable infectivity tissue can be cleaned per manufacturer guidelines.
   6. There is no need to decontaminate or discard the washer-disinfector whose internal components may have been contaminated with prions; use the strictest form of disinfection the machine can tolerate per manufacturer recommendations.

E. **Precautions for Surgical Procedures**
   1. Perform procedure in an operating room.
   2. Involve the minimum required number of healthcare personnel.
   3. Use SINGLE-USE personal protection equipment as follows:
      a. Fluid resistant operating gown, over a plastic apron
      b. Gloves
      c. Mask
      d. Visor or goggles
      e. Linens and covers
   4. Cover all non-disposable equipment.
   5. Maintain one-way flow of instruments.
   6. Use disposable tissue biopsy kit.
   7. Incinerate all disposable items including used sharp containers.
      a. Call SPD or Housekeeping for 2 biohazard bins lined with yellow bag.
      b. Yellow liners/bags are an indication for incineration.
      c. Place used sharp containers along with other disposable items into biohazard bins within yellow bag.
   8. Package & send specimen to the National Prion Disease Pathology Surveillance Center
   9. Quarantine or dispose of instruments (see Appendix A)
   10. **Contact Network Hazmat Safety Officer to coordinate environmental decontamination** (see Appendix B)

Lab

A. Laboratory will not perform routine CSF specimen processing or testing on cases of suspected or known CJD or other prion diseases.
B. Pathology will create a surgical accession number if the specimen requires pathologist assessment when lab/pathology receives a request for paperwork and specimen packaging.
C. Diagnostic reports received from the National Prion Disease Pathology Surveillance Center will be copied and given to the lab manager to be entered into the Electronic Medical Record.

Pathology

A. Pathology is responsible for delivering the National Prion Disease Pathology Surveillance Center paperwork and packaging materials to the requesting unit or department.
B. It is the policy of the Pathology Department not to perform routine tissue processing or frozen section processing on cases of suspected or known CJD or other prion disease.
C. Pathology will coordinate paperwork & diagnostic reports with the Lab as directed in Lab section above.

Precautions For Handling Deceased Patient

A. 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).
B. Nursing manager or designee shall notify morgue and funeral home of known or suspected prion disease
C. If an autopsy is requested, the family of the deceased will be advised to contact the medical examiner's office or the National Prion Disease Pathology Center autopsy coordinator. http://www.cjdsurveillance.com.
D. On the death of a patient with suspected or confirmed CJD/PAD, remove body from the unit following Standard Precautions.
E. It is recommended that the deceased patient be placed in a sealed body bag prior to moving.
    1. Where the skull is open or there is CSF leakage, and where sutures do not completely control this leaking, the bag should be lined with materials to absorb any fluid.
    2. No special precautions are necessary for burial.

References

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Appendix A
QUARANTINE OF INSTRUMENTS

1. Instruments **MUST** be kept moist until cleaned and decontaminated.
2. Avoid mixing instruments used on NO detectable infectivity tissues with those used on HIGH and/or LOW infectivity tissues.
3. Inventory surgical instruments and secure instruments in a robust, leak-proof container labeled "Prion Precautions".
4. Contact Network Hazmat Safety Officer to coordinate storage and gain access to the locked hazardous materials storage.
    a. Store moistened labeled secured instruments in locked hazardous materials storage.
    b. DO NOT TRANSFER INSTRUMENTS TO STERILIZATION DEPT.
    c. Infection Prevention & Control is responsible for the final release of any quarantined instruments.
5. Infection Prevention will consult with Infectious Disease and the Chief Operating Officer to determine disposition of quarantined instruments.
    d. Released instruments will be cleaned & sterilized in regular manner.
    e. Prion contaminated instruments will be decontaminated per WHO Guidelines or incinerated. http://www.who.int/csr/resources/publications/bse/whocdscsraph2003.pdf
6. Infection Prevention should be consulted if there are any questions regarding cleaning procedures.

Appendix B
ENVIRONMENTAL DECONTAMINATION

1) **Contact Network Hazmat Safety Officer to coordinate environmental decontamination.**
2) Gather and prepare decontamination materials;
    a. Bucket, mop & water
    b. Biohazard bin(s) with yellow liner
    c. Pump up type garden sprayer
    d. Personal Protective Equipment (PPE);
        i. Tyvek suit
        ii. Double nitrile gloves
        iii. Disposable rubber over-boots or reusable rubber boots (to be decontaminated after use)
        iv. Powered Air Purifying Respirator (PAPR) with chlorine cartridges
    e. 2 gallons of diluted bleach (sodium hypochlorite) solution*
    f. Yellow 'chemo waste' disposal bags
    g. Container to package contaminated instruments in
    h. Towels
    i. Goggles
3) Don Tyvek suit, rubber over-boots, double nitrile gloves and goggles then prepare an area to doff PPE later on by laying several towels on the floor just inside the OR suite doors.
4) Pour bleach into pump up garden sprayer and pressurize the unit.
5) Remove goggles and don PAPR.
6) Place non-disposable surgical instruments in storage container if not already done so and seal.
7) Clean noncritical environmental surfaces contaminated with high-risk tissues with a detergent and then spot decontaminate these surfaces with prepared bleach (sodium hypochlorite) solution*. Spray bleach on all surfaces having potential contact with tissues of high infectivity (see **Table 1**), wetting or flooding surfaces thoroughly.
8) Maintain a wet surface contact time of at least 15 minutes, re-wetting if necessary.
9) Empty remaining bleach solution into scrub sink and fill sprayer with fresh water.
10) Spray bleach-covered surfaces with rinse water then mop up and rinse with clean water.
11) Place disposable PPE in biohazard bin with yellow liner. Apply yellow "Chemo/Path Incinerate Only" sticker to outer container, close & place in soiled utility room for pick up.
12) Wipe down PAPR with a water damp cloth before returning to storage. The PAPR should not require any additional decontamination as long as proper HAZMAT hygiene protocols are followed.

*Note: Prepare a 1:5 dilution of household bleach (5.25% - 6.15% sodium hypochlorite) and water (1 part bleach to 4 parts water). Sodium hypochlorite products that make pesticidal claims, such as sanitization or disinfection, must be registered by EPA and labeled with an EPA Registration Number. Be familiar with and observe safety guidelines for working with bleach (sodium hypochlorite). Refer to the SDS on the intranet.

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