RANCHO LOS AMIGOS NATIONAL REHABILITATION CENTER

Infection Prevention and Control

SUBJECT: PRION DISEASE, including Creutzfeldt Jakob Disease (CJD)

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Purpose:

To provide guidelines for the infection prevention related management of suspected or confirmed prion disease, including CJD transmissible spongiform encephalopathy, Variant CJD, fatal familial insomnia (FFI), Gertsmann-Straussler-Scheinker Syndrome (GSS), and/or Kuru.

Background:

Transmissible spongiform encephalopathies (TSE) are progressively fatal diseases of humans and animals caused by prions. Human diseases caused by prions include: Creutzfeldt-Jacob Disease (CJD), variant CJD, Gertsmann-Straussler-Scheinker Syndrome (GSS), Kuru, and fatal familial insomnia (FFI). Prions are unique infectious agents and are generally resistant to standard sterilization methods. Clinically, these diseases result in progressive neurologic deterioration and are invariably fatal. High risk patients include those with a history of dura mater transplantation and patients with a known history of cadaver-derived pituitary hormone injection. Currently there is no definitive test to detect an immunologic response to infection although the National Prion Disease Pathology Surveillance Center can test for protein 14-3-3 which serves as a marker for some prion diseases. Variant CJD is caused by a similar prion and for the purposes of this policy, cases of suspected or diagnosed variant CJD should be managed identically to CJD, as should other prior associated diseases. A definitive diagnosis requires a histological examination of the affected brain tissue.

A. Transmission Risk

Transmission of CJD in healthcare settings has rarely been associated with contact to contaminated tissues, specifically central nervous system tissue and eyes as well the use of contaminated neurosurgical instruments or use of infected dura mater, cadaveric growth hormone, contaminated corneal transplants, contaminated electrode implants, and contaminated dura mater grafts. Central Nervous System tissues are considered infectious throughout the symptomatic illness. In most cases, the exact mode of transmission in humans is not known. Potentially infectious body fluids and tissue likely carry the most risk of transmission during invasive procedures. Transmission of CJD has not been associated with environmental contamination, fomites, or person-to-person via skin contamination.

The World Health Organization (WHO) categorizes potentially infectious body fluids and tissues into three risk groups. The following list identifies which body fluids and tissues are potentially infectious:

INFECTIVITY	SUBSTANCES
High Infectivity Tissue	Central Nervous System, specifically: Brain (including dura mater), Spinal Cord, Posterior Eye, Pituitary tissue
Low Infectivity Tissues and Fluids	Cerebrospinal fluid, Kidney, Liver, Lung, Lymph nodes, Spleen, Placenta and Olfactory epithelium (vCJD can be found in lymphoid tissue throughout the body, including tonsils and intestines)
No Detectable Infectivity	Blood, Leukocytes, Serum, Adipose, Adrenal gland, Gingival tissue, Heart muscle, Intestine, Peripheral nerve, Prostate, Skeletal muscle, Testis, Thyroid gland, Tears, Nasal mucous, Saliva, Sputum, Sweat, Serous exudates, Breast Milk, Semen, Urine, Feces, Bone Marrow, Vaginal Secretions

B. Public Health Reporting of CJD

CJD is a reportable disease in the United States and must be reported to the Los Angeles County Department of Public Health within 7 calendar days. See section “C. Notification Procedures” for details on hospitalized patients.

C. Notification Procedures

1. Attending Physician and/or Admitting Physician: It is the responsibility of the attending and/or admitting physician to inform the following departments/staff upon admission of a CJD patient, or at the time that a diagnosis of suspected or confirmed CJD is being contemplated:

   i. Infectious Disease
   ii. Infection Prevention and Control Department
   iii. Admitting Area Nurse Manager or Charge Nurse
   iv. Department of Pathology
   v. House Nursing Supervisor after normal business hours, weekends and holidays

2. Prior to scheduling a patient with suspected or confirmed CJD for any invasive procedure (i.e. lumbar puncture), the area Charge Nurse and the area Nurse Manager must be notified. **Note: All CSF specimens obtained in addition to the usual patient identification information, must be labeled as a "biohazard" and as "Suspected CJD" or "Suspected Creutzfeldt-Jacob Disease".**
3. If an operating surgeon believes that a patient is at risk for CJD, he or she must communicate that information to:

   i. Operating Room (OR) Nurse Manager or Charge Nurse
   ii. Anesthesiology staff
   iii. Neuropathology or Clinical Laboratory Administrative Coordinator/Lab Director
   iv. Risk Management
   v. Infection Prevention and Control Department

4. Contaminated items that have not been processed appropriately must be recalled (e.g. medical devices used for brain biopsy before diagnosis). Pathology department must report all prion positive specimens to the Infection Prevention and Control Department who will assess the incident and notify the appropriate departments (eg, OR, Risk Management).

Control Measures:

Whenever possible, performing neurosurgical procedures on a patient with suspected CJD should be avoided. All suspected CJD cases that are scheduled for brain biopsies will be managed according to the following guidelines, which are intended to reduce the risk of contamination of the surgical instruments and possible exposure to prion disease.

Instruments used for biopsy of possibly infective tissue with no confirmed diagnosis should be quarantined until confirmation of disease, or cleaned and sterilized according to CJD protocol.

Because the prion is difficult to eradicate, stringent sterilization precautions must be taken with all surgical instruments.

A. General Infection Prevention Precautions
   1. Meticulous attention to Standard Precautions should be used in the care of all patients, in the handling of tissue samples or specimens, and in the decontamination of equipment/work surfaces exposed to known or suspected prion disease.
   2. An isolation room is not necessary.
   3. Any samples and specimens must be labeled as a "biohazard" and as "Suspected CJD" or "Suspected Creutzfeldt-Jacob Disease" in addition to the usual patient identification before being sent to the laboratory.

4. Tissues should be fixed with formalin.
5. Tag equipment that requires special prion reprocessing after use.
6. Restrict OR traffic and keep supplies to a minimum to facilitate containment and cleanup. The OR staff will be responsible for the post procedure cleanup.
7. Instruments/supplies that require a method of sterilization other than steam sterilization must not be used (eg, flexibile endoscopes) or if used, must be discarded.
8. Manual equipment (eg, saws, drills) should be used rather than power equipment due to the impossibility of adequately cleaning and sterilizing power equipment without destroying it.
9. Neurosurgical instruments used on these cases should be disposable.

B. Medical Item and Environmental Surface Decontamination

A component that should be integrated into the disinfection and sterilization process is the risk of infection associated with the use of the medical device. There are three risk categories of medical devices that can be assigned: critical, semi-critical, or non-critical.

Critical Items enter sterile tissue or the vascular system (eg, surgical instruments and implants)

1. Critical items that have been contaminated with high-to-low risk tissues must be reprocessed with the following procedure (steps i. through iii.):
    i. Instruments should be kept wet (eg, immersed in water or prioncidal detergent) or damp after use and until they are decontaminated.
    ii. The instruments should be decontaminated as soon as possible after use (eg, high level disinfection).
    iii. After decontamination, the items must be sterilized, one of the following options must be completed:

    a. Option 1 - Autoclave 134ºC for 18 minutes in a pre vacuum sterilizer.
    b. Option 2 - Autoclave 132ºC for 1 hour in a gravity displacement sterilizer.
    c. Option 3 - Immerse in 1N NaOH (1N NaOH is solution of 40g NaOH in 1L water) for 1 hour; remove and rinse in water, then transfer to an open pan and autoclave (121ºC gravity displacement sterilizer or 134ºC porous or prevacuum sterilizer) for 1 hour.
    d. Option 4 - Immerse in 1N NaOH for 1 hour and autoclave in gravity displacement sterilizer at 121ºC for 30 minutes, then clean and subject to routine sterilization.

2. Critical items that have been contaminated with no-risk tissues must be reprocessed with the following procedure:
    i. Decontaminate and sterilize these devices using conventional protocols of heat or chemical sterilization.
    ii. Use standard cleaning and high level disinfection protocols for reprocessing rigid endoscopes (except neurosurgical endoscopes with central nervous

system contact which require processing as described above).

Semi-Critical items come into contact with mucous membranes or non intact skin (eg, some endoscopes and respiratory therapy equipment)

1. Follow procedures for the reprocessing of critical items.
2. Flexible endoscopes cannot be adequately disinfected after exposure to patients with suspected or known CJD. Other alternatives to flexible endoscopy should be considered. Therefore, it is recommended if absolutely needed, that a flexible endoscope at the end of its useful life be used. The flexible endoscope will have to be disposed of afterwards.

Non-Critical items come into contact with intact skin and non-mucous membranes (eg, floors, walls, blood pressure cuffs, and furniture)

1. Non-critical environmental surfaces contaminated with high- and low-risk tissues (e.g. a laboratory surface in contact with brain tissue of a CJD-infected person):
    i. Clean these surfaces with a detergent, then spot-decontaminate these surfaces with a 1:5 dilution of sodium hypochlorite (i.e. bleach: a 1:5 dilution of 5.25% - 6.15% sodium hypochlorite provides 10,500 -12,300 ppm chlorine) for a contact time of 15 minutes.
    ii. Use disposable plastic-backed cover sheets on work surfaces.
2. Non-critical equipment contaminated with low- and high-risk tissues:
    i. Clean and then disinfect equipment using a 1:5 to 1:10 dilution of sodium hypochlorite or 1N NaOH, depending on material compatibility.
    ii. Ensure all contaminated surfaces are exposed to the disinfectant.
3. Non-critical environmental surfaces contaminated with low-risk tissues require only standard disinfection.
4. Non-critical equipment and non-critical surfaces contaminated with no-risk tissues or fluids:
    i. Use standard disinfection to process equipment and surfaces.
    ii. Use disinfectants recommended by OSHA for decontaminating blood-contaminated surfaces (e.g. 1:10 to 1:100 dilution of 5.25% - 6.15% sodium hypochlorite)

C. <u>General Guidelines for the Disinfection and Sterilization of Contaminated Medical Devices</u>

1. Always follow manufacturer recommendations before initiating the disinfection and sterilization process described above. Some devices may not be resistant to certain chemicals and/or temperatures and may be damaged irreparably by this process. Prion-contaminated medical devices that are impossible to clean or fully expose to steam and other sterilants must be discarded by incineration.
2. Flash sterilization must not be used for reprocessing.
3. Recall contaminated items (e.g. medical devices used for brain biopsy before diagnosis) that have not been processed appropriately.

D. Quarantine

Quarantine can be used to avoid needless destruction of instruments when suspect cases are later found not to have prion disease. If the department can safely quarantine instruments until a diagnosis is confirmed, the following procedure must be implemented:

1. Items for quarantine should be cleaned by the best non-destructive method as per Section "B. Medical Item and Environmental Surface Decontamination" procedures. The items must be sterilized, packed, dated, "Hazard" labeled, and stored in specially-marked rigid, sealed containers.
2. Monitoring and ensuring maintenance of quarantine is essential to avoid accidental re-introduction of these instruments into the circulating instrument pool.
3. If prion disease is excluded as a diagnosis, the instruments may be returned to circulation after inspection for proper function and appropriate sterilization.

E. Waste Disposal

1. All disposable instruments, materials, supplies contacting spinal fluid and other regulated medical waste (eg, bulk blood, pathologic waste) must be incinerated.
2. Items used on these patients should be disposable, if possible, and placed in red bags prior to leaving the OR.
3. Needles, scalpels, and other sharps must be disposed of immediately in the sharps disposal box. The safety device must be activated at the time of disposal.

F. Handling deceased patients

1. It is recommended the deceased patient be placed in a sealed body bag prior to moving. The bag should be lined with materials to absorb any fluid if the skull is open, there is CSF leakage, and if sutures do not completely control leakage.
2. When a patient dies, the morgue and funeral home must be notified that the patient had prion disease/CJD. No excess precautions need to be taken with regard to burial (e.g. no special cemetery is required).

G. Conducting an Autopsy

1. Disposable protective clothing should be worn, including surgical cap and gown, apron, double gloves, and a face visor which completely encloses the operator's head to protect the eyes, nose, and mouth. Consider the use of armored or cut-resistant gloves for hand protection.
2. Disposable or dedicated reusable instruments are recommended in order to minimize the risk of environmental contamination.
3. See section B. Medical Item and Environmental Surface Decontamination and section C. General Guidelines for the Disinfection and Sterilization of Contaminated Medical Devices for instrument processing after an autopsy.

H. Occupational Exposures The theoretical risk results from exposure to high-infectivity tissue through needlestick injuries with inoculation.
   1. A non-percutaneous exposure to blood and body fluids of CJD patients should be followed by a thorough washing with soap and water (avoid scrubbing), rinse, and drying. For maximum safety, consider a brief exposure to 0.1 N NaOH or a 1:10 dilution of bleach for 1 minute.
   2. Needle sticks or lacerations: gently encourage bleeding. Wash (avoid scrubbing), rinse, and dry. For maximum safety, rinse wound with 0.5% sodium hypochlorite and then rinse with water.
   3. Splashes into the eye or mouth: irrigate with either saline (eye) or tap water (mouth) for several minutes.
   4. All possible exposures to CJD must be reported to Employee Health.

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