

INFECTION CONTROL POLICIES AND PROCEDURES FOR PATIENTS WITH SUSPECTED OR CONFIRMED HUMAN PRION DISEASE (e.g., Creutzfeldt-Jakob Disease [CJD])*

**This is a living document which will be updated as more information becomes known.*

I. PURPOSE:

To define precautions for handling materials from any patient with suspected or confirmed prion disease and transmissible spongiform encephalopathy (TSE). At UCSF, the most commonly-seen human prion diseases include: sporadic Creutzfeldt-Jakob disease (sCJD), familial Creutzfeldt-Jakob disease (fCJD), and Gertsman-Straussler-Scheinker disease (GSS).

II. OBJECTIVES:

- A. To provide basic background on human prion diseases
- B. To prevent transmission of prions to personnel, other patients or the community
- C. To confine, contain, and safely handle contaminated materials generated during the course of hospitalization
- D. To define contaminated materials and tissues and describe their appropriate disposal

III. BACKGROUND INFORMATION:

Transmissible spongiform encephalopathies (TSEs), or prion diseases, are inevitably fatal neurodegenerative brain and central nervous system diseases affecting humans and animals. Prion diseases affecting humans can be divided into three categories:

- A. Spontaneous or Sporadic (sCJD)
 - Sporadic CJD
 - Sporadic fatal insomnia (<10 cases known in world history)
- B. Genetic or Familial
 - Familial CJD
 - Familial fatal insomnia (FFI)
 - Gertsman-Straussler-Scheinker disease (GSS)
- C. Acquired
 - Iatrogenic CJD (iCJD)
 - Variant CJD (vCJD) (also called new variant or nvCJD)
 - Kuru (extinct)

IV. EPIDEMIOLOGY

About 85% of human prion disease cases are sporadic, usually sCJD, with no recognizable pattern of transmission. 10% - 15% of human prion diseases are genetic, and less than 1% is acquired. The incubation period for sporadic and genetic cases is unknown. Cases of iCJD, particularly those with defined exposure events to high-risk tissue, appear to develop symptoms between 1.5 years to >2 decades after exposure. Human prion disease is present worldwide with an incidence of about 1 case per million people each year. In the U.S., there are approximately 250-400 cases per year, and about 20 cases per year in California.

sCJD typically affects people between 55 and 75 years of age. Death in CJD patients occurs within 6-12 months post onset and is often due to pneumonia or other complicating conditions of dementia. There is no proven treatment or prophylaxis. However, treatment studies are underway. (See <http://memory.ucsf.edu/Research/MAC.htm>)

Variant Creutzfeldt-Jakob disease (vCJD) was diagnosed in the United Kingdom in 1996. Since then, there have been >150 cases of vCJD, almost all occurring in the U.K. However, as of June 2005, the following countries have reported cases: France, Italy, Portugal, Oman, Japan, Republic of Ireland,

Canada, and Hong Kong. No cases of vCJD have been acquired in the western hemisphere; one patient in Florida (U.S.) and one patient in Canada died of vCJD, but both lived in the U.K. during the peak of the epidemic, and are presumed to have acquired the disease there.

Evidence suggests that vCJD is acquired through ingestion of bovine spongiform encephalopathy (BSE, or "mad cow disease")-tainted meat. Two cases of confirmed BSE in domestic cows have been identified in the U. S. as of June 2005. The first case, in December 2003, was from a cow imported from Canada. The second animal, killed in November 2004 and confirmed with BSE in June 2005, is believed to have originated in the U.S. It is unclear whether BSE is a food borne hazard in the U.S. Although CJD is not a reportable disease in most states, the CDC monitors incidence of CJD in the U.S. While the U.S. Department of Agriculture has conducted active surveillance efforts for BSE since May 1990, there is controversy about whether these efforts are adequate.

V. HUMAN PRION DISEASE SIGNS AND SYMPTOMS

sCJD is the most common form of human prion disease, characterized by rapidly progressive dementia, motor dysfunction, visual symptoms (e.g., double vision, visual misperception and distortion), behavioral changes, myoclonus, and a variety of other neurologic signs and symptoms. MRI often reveals classic abnormalities in sCJD as well as in some genetic and acquired forms of human prion disease. Electroencephalograms may show characteristic periodic sharp waves, but often not until later stages of the disease.

Genetic forms of human prion disease can present exactly like sCJD with rapid onset of neurological symptoms, classic MRI findings, and death within months of onset. Genetic forms of human prion disease can also present as slowly progressive ataxia or Parkinsonian illness progressing over years. In about 60% of genetic human prion disease, there is no known family history, although careful family history will often reveal previously misdiagnosed Parkinson's or Alzheimer's disease.

vCJD may be more readily transmitted than other forms of human prion disease. vCJD typically presents with a prodrome of several months' duration characterized by profound psychotic illness, followed by onset of neurologic disease. Neurologic symptoms often include movement disorders (dystonia, myoclonus and ataxia), persistent painful sensory symptoms, and dementia.

For more information about human prion disease, view <http://memory.ucsf.edu>

VI. CHARACTERISTICS OF PRIONS

The causative agents of TSEs are prions (*prēe-ahns*), small protease-resistant proteins which are resistant to the conventional methods of sterilization and decontamination. However, because prions are proteins, they can usually be inactivated by procedures that denature or hydrolyze proteins such as exposure to sodium hydroxide or autoclaving at very high temperatures for prolonged periods of time. If prions come into contact with certain materials, such as metal and glass, they can be much more difficult to inactivate.

VII. DISTRIBUTION OF PRIONS IN HUMANS

Infected brain tissue, dura mater, spinal cord, and cornea are highly infectious (see table below for presumed infectious risk of human tissues and fluids). Due to the hardiness of the causative agent, special precautions must be taken when handling items that have come into contact with these highly infectious tissues. Although cerebral spinal fluid (CSF) has not been shown to readily transmit CJD, because of its contact with highly infectious tissues, at UCSF it has been decided to err on the side of caution for handling CSF. Procedures for handling CSF are detailed in Table 3.

VIII. DISTRIBUTION OF INFECTIVITY OF PRIONS IN THE HUMAN BODY

Table 1. Distribution of infectivity of prions in the human body

INFECTIVITY CATEGORY	TISSUE, SECRETION, EXCRETION		
HIGH INFECTIVITY or HIGH RISK (Listed alphabetically)	brain (including dura mater), spinal cord, cranial ganglia cranial nerves eye, e.g., cornea (<i>invasive procedures, e.g., intravitreal injections, ocular surgery</i>)		lymphoreticular tissues (<i>e.g., tonsils, adenoids, appendix</i>)*** spinal cord spinal ganglia
MEDIUM or LOW INFECTIVITY or MEDIUM or LOW RISK ** (Listed alphabetically)	adipose tissue adrenal gland blood* bone marrow CSF dental pulp feces gingival tissue heart intestine kidney liver lung	lymph node muscle nasal mucous olfactory epithelium peripheral nerve peripheral nerves placenta prostate sclera (<i>non-invasive eye procedures involving contact with superficial corneal epithelium, sclera, conjunctiva [e.g., tonometry, gonioscopy, ERG]</i>) serous exudate	skeletal muscle spleen spleen testis thymus thyroid gland
INSUFFICIENT DATA TO DETERMINE INFECTIVITY or NO INFECTIVITY (Listed alphabetically)	milk saliva semen sputum		sweat tears urine

*Only vCJD has been shown to be transmissible by blood from human to human, and that only through blood transfusion of donated blood. It is not known what the minimum amount of vCJD-contaminated blood is required for transmission. Although transmission from blood in other human prion diseases (not vCJD) has not been shown, there is a theoretical risk; therefore any exposure to blood from patients with confirmed or suspected prion disease must be considered exposure to prion disease.

**Some tissues in this section have not yet been shown to contain prions, or have not yet been shown to transmit disease, but have been placed in this category because of the blood exposure involved in sampling the tissues.

***Lymphoreticular tissue in vCJD is considered high risk

IX. MODES OF TRANSMISSION

The transmissibility of prions has been demonstrated by inducing disease in laboratory animals. The most effective method of infection of animals was by intracerebral inoculation of prions, with intraperitoneal and percutaneous inoculation being significantly less effective, and ingestion of prions the least effective.

- A. *Iatrogenic*: Iatrogenic transmission is exceedingly rare (313 cases total as of June 2005). It has occurred after transplantation of CJD-infected corneas (3 cases), dura mater grafts (>136 cases), and stereotactic depth electrodes previously used on infected individuals (3 cases). Iatrogenic CJD has been described in patients who received injections of human pituitary-derived hormones (168 cases).

- B. *Occupational:* **There is no evidence of occupational transmission of CJD to healthcare workers.** Although cases of human prion disease have been reported in approximately 24 healthcare workers, this incidence does not exceed what would be expected by chance alone. The highest theoretical risk is from occupational exposure to high infectivity tissue through needlestick, splashing of the mucous membranes or unintentional ingestion. All healthcare personnel who work with patients with known or suspected prion diseases must adhere to standard precautions.

TRANSMISSION OF CJD HAS NOT BEEN ASSOCIATED WITH ENVIRONMENTAL CONTAMINATION OR FOMITES.


X. ROUTINE CARE OF PATIENTS


Normal social (including intimate) and clinical contact and non-invasive clinical investigations (e.g., imaging, EKG, EEG) of human prion disease patients do not present a known risk to healthcare workers, visitors, or the community. Based on current knowledge, isolation of patients is not necessary; they can be cared for using Standard Precautions (See Infection Control Manual). A private room is not required for infection control purposes. Patient waste will be handled in accordance with hospital policy (Infection Control Manual Section, Environment of Care Manual). Liquid body substances such as blood, urine, bile, vomitus, or other secretions/excretions, and stool are disposed of in the sanitary sewer (toilet, hopper, or lab sink) in a manner that limits splash. Feeding utensils, feeding tubes, suction tubes, or items used in skin or wound care, and bed linens are handled as per routine.

XI. PROCEDURES/AREAS REQUIRING ADDITIONAL INFECTION CONTROL PRECAUTIONS due to potential exposure to low or high infectivity fluids or tissues.

- A. Lumbar Puncture in a patient in whom Human Prion Disease is confirmed or is in the differential diagnosis.

Table 2. Steps in performing bedside lumbar puncture, and steps in room decontamination

Steps in Procedure	Comments
<p>1. Scheduling of Lumbar Puncture (LP) Procedure</p> <ul style="list-style-type: none"> a. The patient will be in a procedure room or a room with roommate absent during a lumbar puncture procedure. b. Order a CJD Lumbar Puncture Kit that contains the supplies needed for the procedure from Material Services. c. Post a sign on the door of the room to indicate "Procedure in Progress Do not Enter" d. All personnel assisting with the procedure must wear personal protective equipment. 	<p>To prevent other personnel from entering during the procedure.</p>
<p>2. Equipment & Supplies for LP</p> <ul style="list-style-type: none"> a. The CJD Lumbar Puncture Kit contains the following supplies: <ul style="list-style-type: none"> • Infection Control Procedure for Patients with CJD • Disposable Lumbar Puncture Tray • 4 long-sleeved fluid repellent disposable gowns • 4 masks with face shield • 2 alternative eye protection goggles • 2 face masks without face shield • Signage for patient room "Procedure in Progress Do Not Enter" • One quart size sharps container • Occlusive dressing material to cover LP site: 2 sterile 2x2's and 2 (two) 10 cm x 8 cm transparent dressings • 3 pw chemical hazardous waste bags for disposal materials contaminated with any materials used during the LP procedure. • 3 OEH&S chemical pickup request forms and tags with instructions and phone numbers. b. Cover immediate exposed bed, linen, and bedside table with disposable impermeable barriers. c. Set up a sterile field next to patient with a disposable impermeable barrier. d. Place a sharps container in close proximity to the 	<p>To prevent contamination of other work surfaces with CSF</p> <p>To prevent contamination of surfaces with CSF</p>

Steps in Procedure	Comments
<p>LP procedure area. The used lumbar puncture needle and all sharps used during the procedure shall be discarded in this container. This container must be placed in the yellow chemical hazardous waste bag.</p>	
<p>3. Personal Protective Equipment All health care workers performing or assisting with the procedure, who may come in contact with CSF, must wear the following personal protective equipment:</p> <ol style="list-style-type: none"> Long sleeve fluid repellent gown Face protection to include eyes, nose and mouth protection (mask with face shield). Gloves. MD performing the procedure will wear double sterile gloves. 	
<p>4. Transport of specimen(s) to the lab</p> <ol style="list-style-type: none"> CSF may be infectious and must be handled with care. CSF will be double-bagged in leak-proof specimen bags and labeled "CJD Precautions." Document where specimen will be delivered, who will be transporting the specimen, and notify the lab receiving the specimen that it has been collected. Lab personnel shall use universal precautions when handling specimens (see laboratory procedures below). 	
<p>5. Post Procedure Cleaning:</p> <ol style="list-style-type: none"> <u>Any person in the room may identify a potentially contaminated CSF spill, and all have equal responsibility to order a contaminated clean-up procedure</u> Obtain PSA Clean-up Kit, consisting of protective equipment wrapped separately, consisting of <ul style="list-style-type: none"> 2 pairs blue high-risk gloves, 2 fluid repellent gowns, 2 masks with face shield, 2 pairs of fluid repellent shoe and leg covers. Remove patient's gown if contaminated with CSF during the procedure and discard in yellow chemical hazardous waste bags (not to be confused with yellow personal effects bags). Discard any dressing used to cover the lumbar puncture site in yellow chemical hazardous waste bag. Discard the sharps container and all 	<p>A CJD Spill Tote* may be ordered from  erial Services.</p> <p>Personnel must be trained and observe safety guidelines when working with 1N sodium hydroxide. With the recommended precautions, it can be used safely.</p> <p>Sodium hydroxide must not come in contact with strong acids, flammable liquids and solvents, e.g., acetone.</p> <p>Linen that has not been contaminated with CSF during the procedure can be placed in the covered linen container for usual reprocessing</p> <p>If no CSF contamination has occurred, standard cleaning is appropriate. Any person in the room may identify a potentially contaminated CSF spill, and all have equal responsibility to order a contaminated clean-up procedure.</p>

Steps in Procedure	Comments
<p>disposable items into yellow chemical hazardous waste bag.</p> <p>e. Remove disposable barriers covering the linen. Discard any linen contaminated with CSF in the yellow chemical hazardous waste bag.</p> <p>f. Discard all yellow chemical hazardous waste bags into the yellow barrel labeled CJD WASTE ONLY for pickup and incineration.</p> <p>g. PSA's will wear fluid repellent gowns, eye, nose and mouth protection, leg and shoe covers, and blue high-risk gloves for cleaning the room.</p> <p>h. If CSF contamination is present, use 1N sodium hydroxide to wipe down all surfaces that have come in contact with the CSF, including the bed, bed rails and bedside table. 1N sodium hydroxide will be poured onto rags to damp wipe. Damp wipe area generously with the 1N sodium hydroxide solution and let sit undisturbed for one hour.</p> <p>i. If CSF fluid has splashed onto floor, clean floor by carefully pouring small amounts of 1N sodium hydroxide directly onto floor at floor level, taking care not to splash.</p> <p>j. After one hour, rinse area thoroughly with water then clean the floor and surrounding area according to standard procedure.</p> <p>k. Unused 1N sodium hydroxide will be tightly capped and double bagged into yellow hazardous waste bags.</p> <p>l. Complete and affix Hazardous Waste tag and place into receptacle provided for liquid waste.</p> <p>m. Double bag all cleaning equipment, e.g., rags and mop heads that have come in contact with 1N sodium hydroxide into separate yellow hazardous waste bags.</p> <p>n. Affix Hazardous Waste tag(s) to each bag and complete and fax OEH&S chemical pickup request forms for solid and liquid waste per instructions on form.</p> <p>o. Place sharps container in a yellow chemical hazardous waste bag.</p> <p>p. Place tagged yellow bags in the designated yellow barrel labeled CJD WASTE ONLY in a secured area until picked up by OEH&S.</p>	<p>1N sodium hydroxide is available from Material Services or, in an emergency, Central Sterile Processing.</p> <p>To prevent creating breathing hazard, do not use spray bottles.</p> <p>Do not use mop bucket.</p> <p>Personal protective equipment includes:</p> <ul style="list-style-type: none"> • fluid repellent gown • face shield/goggles • blue high-risk gloves • shoe and leg covers if shoe contamination is possible <p>Designated barrels are located in ACC, OR, 8th floor, GCRC</p>
<p>6. Procedures for sodium hydroxide splash and spills onto eyes, clothing or skin</p> <p>a. Splashes to the eye must be treated immediately!</p> <p>Rinse affected eye(s) thoroughly for 15 minutes with water or normal saline. Do not rub or keep eyes closed. After initial rinse,</p>	<p><u>DO NOT TREAT EYE EXPOSURES WITH SODIUM HYDROXIDE OR BLEACH</u></p> <p>Aerosolized exposure to sodium hydroxide may cause lacrimation (tearing), blurred vision, and photophobia as well as chemical conjunctivitis and</p>

Steps in Procedure	Comments
<p>notify the Needlestick Hotline for evaluation.</p> <p>b. If sodium hydroxide is splashed or spilled onto the skin or clothing, rinse skin and affected clothing thoroughly with water for 5-10 minutes.</p>	<p>corneal damage. Bring the 1N sodium hydroxide MSDS to the ER.</p> <p>Sodium hydroxide is caustic but fortunately it is slow-acting and can be rinsed out with water. If left on skin or clothing for an extended time, 1N sodium hydroxide will eventually cause a skin burn. Burn may appear after an aching sensation, and in milder cases a skin rash may appear. Other symptoms are cold and clammy skin with cyanosis or pale color.</p>

*CJD Spill Tote contains:

- One copy of this policy
- Three fluid repellent gowns
- Three masks with face shields
- Three alternative eye protection goggles and masks
- Blue high-risk gloves (2 medium and 1 large)
- Three pairs of fluid repellent leg and shoe covers
- Six yellow chemical hazardous bags
- Six OEH&S chemical tags
- Six OEH&S chemical pickup request forms with instructions and phone numbers
- Two liters 1N sodium hydroxide
- Two liters 2N sodium hydroxide (for use in suction canisters)
- One box extra strength shop rags
- One mop head
- MSDS sheet for sodium hydroxide

B. Clinical Laboratory

All CSF specimens will be considered potentially infectious for CJD and will be handled the same as CSF specimens from patients known or suspected to be infected with CJD.

Table 3. Clinical Laboratory Procedures for handling fluids potentially contaminated with prions

Steps in Procedure	Comments
<p>1. CSF</p> <p>a. All staff will wear Personal Protective Equipment when handling CSF specimens.</p> <p>b. In each section, a work area will be defined for handling CSF specimens. The area will be covered with an absorbent barrier drape, which will be changed once per shift or after a CSF spill.</p> <p>c. All CSF specimens will be spun down in disposable, capped containers. Specimens received in Central Processing will be spun down in the original plastic, capped collection tube, then distributed in capped tubes to the sections (Chemistry, Immunology) or poured off for Send Out. Specimens received in Hematology and Microbiology will be pipetted into disposable plastic funnels, capped and spun down in the cytocentrifuge in each section. If a sample is re-spun in Immunology, it will be spun down in a capped, disposable tube or funnel.</p> <p>d. Used CSF funnels and collection tubes, and all remaining CSF</p>	<p>At a minimum, staff will wear gloves and lab coats at all times when working with CSF specimens and slides. Where the potential for splash exists, face shields (protective eye wear and mucosal protection) will also be worn.</p>

Steps in Procedure	Comments
<p>after testing is completed (including from patients with known or suspected CJD) will be placed in the pathological waste tub. .</p> <p>e. All equipment (e.g. inside cover of cytocentrifuge) and work areas used to process CSF specimens will be wiped down once every shift with 10% bleach and allowed to dry for 10 minutes. The work area will be re-covered with a barrier drape.</p> <p>f. Reusable supplies (e.g. counting chambers) will be soaked in full strength household bleach for one hour after use on CSF specimens.</p> <p>g. Each section will process CSF specimens as follows:</p> <ol style="list-style-type: none"> 1. Chemistry will pour CSF into disposable cup and run sample through automated analyzers. Sample will then be discarded in regulated pathological waste. When required, unused CSF will be returned to Central Processing for distribution to Immunology. 2. Immunology: CSF specimens will be pipetted into open disposable well of automated analyzer. After testing, the specimen will be discarded in regulated pathological waste. CSF specimens will also be placed onto non-disposable slides, rotated (8 minutes) and then read on the CSF microscope. The non-disposable slides will be soaked in full strength bleach for one hour. The barrier drape on the rotator surface will be changed after each use and the rotator lid wiped down with a 10% bleach towelette and air dried for 10 minutes. The CSF microscope will be wiped down according to manufacturer's recommendations. 3. Hematology: CSF specimens will be pipetted onto a counting chamber and read. After cytocentrifugation, all CSF slides will be processed on the stainer. Microscopes used to review CSF slides will be wiped down according to manufacturer's recommendations. All non-disposable supplies (e.g. counting chambers) will be soaked in full strength household bleach for 1 hour after use on CSF specimens. 4. Microbiology: The areas for handling CSF specimens will be covered with an absorbent barrier drape. Spun CSF specimens will be pipetted onto a slide, which will be stained and dried on the heating block. The slide will then be read on one of three microscopes and stored with the Microbiology slides. Gloves will be worn when using the microscopes or when handling CSF slides. Disposable supplies will be used when plating CSF specimens and the supplies discarded in the pathological waste tubs after use. The following work areas will be wiped down with 10% household bleach once per 	<p>Gloves will be worn when handling slides, or working with the stainer.</p> <p>Due to the need to wipe down microscopes, the number of scopes used to review CSF slides will be kept to a minimum.</p>

Steps in Procedure	Comments
<p>shift or after a CSF spill and the absorbent barrier drape changed: planting biological safety cabinet, antigen testing area (work surface of plexiglass shield, heating block, slide rotator surface, and rotator lid), work area adjacent to the microscope for reading CSF smears, and PCR biological safety cabinet in pre-PCR room. The microscope will be wiped down according to manufacturer's recommendations. Non-disposable supplies, such as Cryptococcus antigen latex agglutination slides, will be soaked in full strength bleach for one hour after use with CSF specimens.</p> <p>h. CSF Spills:</p> <ol style="list-style-type: none"> 1. After any CSF spill, the area of contamination will be wiped down. If the spill is contained on the barrier drape, the work area and equipment (where practical) will be wiped down with 10% household bleach and air dried for 10 minutes. If the spill occurs other than on the barrier drape, the area will be flooded with full strength household bleach and allowed to air dry. 2. Materials used to clean the spill (cloth or paper towel, absorbent barrier etc.) will be placed in the regulated pathological waste tub 3. If an exposure occurs, refer to section XV, Occupational Exposure. 4. Do not use bleach in the eyes or mouth. For a splash to the eye or mucous membrane, rinse well with water or saline. 5. If lab coat or personal clothing is contaminated with the splash, remove garment and place in regulated pathological waste. 6. Immediately call the Needlestick Hotline @ 719-3898 to report an exposure. 	<p>One pair each of medium and large scrubs will be available in the Chemical Spill Materials cabinet in L554 (Chemistry).</p>

Table 4. Handling tissues in Pathology

Steps in procedure	Comments
<p>2. Tissue: All tissue samples identified as "high," "medium" or "low" infectivity or risk (see Table 1) will be treated as potentially infectious and will be handled as follows.</p> <ol style="list-style-type: none"> a. All staff will wear Personal Protective Equipment when handling tissue specimens. At a minimum, staff will wear gloves and a lab coat at all times when working with these specimens. In conditions under which aerosolization may occur, face shields (protective eye wear and mucosal protection) will also be worn. b. The work area for these tissues will be defined and protected by a disposable barrier drape. c. When possible, disposable supplies will be used in the preparation of these tissue samples and will be discarded in the pathological waste tubs. Sharps used in the preparation of specimens will be discarded in an approved sharps container and then into the pathological waste tub. d. After testing, tissue specimens from all sections will be placed in 	

Steps in procedure	Comments
<p>1N NaOH for 1 hour. The tissue will be removed from the NaOH and autoclaved at 134 degrees C for 1 hour in the Microbiology autoclave. Caution must be used when removing specimens from the autoclave, for disposal in the pathological waste tubs.</p> <p>e. Decontamination after Tissue Specimen Preparation</p> <ol style="list-style-type: none"> 1. After processing the specimen, the barrier drape will be removed and discarded in the pathological waste tubs. The work area and all equipment (when feasible) will be wiped down with 10% household bleach and allowed to air dry. 2. Non-disposable supplies (such as tissue grinders) will be soaked in full strength bleach for one hour. <p>f. Contamination of the work area:</p> <ol style="list-style-type: none"> 1. Should contamination of the work area occur, the barrier drape will be discarded in the pathological waste tubs, and the area flooded with full strength household bleach and allowed to air dry. 2. Materials used to clean the spill (cloth or paper towel, absorbent barrier etc.) will be placed in the pathological waste tubs. 3. If an exposure occurs (skin contact, laceration and/or puncture) the affected area will be washed for 2-3 minutes with 10% Household bleach, followed by copious amounts of water. Cover any wound with a waterproof dressing. 4. Do not use bleach in the eyes or mouth. For a splash to the eye or mucous membrane, rinse well with water or saline. 5. If lab coat or personal clothing is contaminated with the tissue, remove garment and place in pathological waste tubs. One pair each of medium and large scrubs will be available in the Chemical Spill Materials cabinet in L554 (Chemistry). 6. Immediately call the Needlestick Hotline @ 719-3898 to report an exposure. 	

Table 5. Patient handling in Imaging

Steps in Procedure	Comments
<p>C. MRI</p> <p>Schedule no later than 12:00 noon with a latest time for entering the magnet of 2:00 PM.</p> <ol style="list-style-type: none"> A. An attending anesthesiologist, a registered nurse from Radiology and an anesthesia workroom technician will be present at induction of anesthesia or sedation. B. Five minutes before the end of the scan, the MRI technologist will call a Radiology RN and anesthesia workroom technician to be present when the patient comes out of the scanner and until the patient is transferred to the receiving department. C. Anesthesia equipment or supplies will be disposed of in accordance with recommended anesthesia procedures. (see VIII.B. Anesthesia) 	<p>Need to add procedures for room cleaning, other procedures that might be done in imaging department (such as fluoroscopy-guided LP)</p>

D. PERIOPERATIVE CARE FOR PATIENTS WITH SUSPECTED OR CONFIRMED HUMAN PRION DISEASE

All patients with a diagnosis of suspected or confirmed CJD require a higher level of infection control precautions in the Operating Room. Surgeons (attending or resident) booking a case must document all suspected or confirmed CJD cases on the Emergency Scheduling slip. If the diagnosis is not documented, the scheduling coordinator will contact the surgeon for clarification. For example, if a Burr hole procedure is booked for 'Brain Biopsy: other' (i.e., not for tumor), then it must be clarified with the Neurosurgery service that CJD is not on the differential. **The scheduling coordinator shall notify the charge nurse immediately if a tonsil biopsy or procedure not involving the neurosurgical service is ordered on a CJD patient. Cases should be scheduled early in the day to allow adequate time for cleaning and processing of specimens in surgical pathology.**

1. PRE- OPERATIVE PROCEDURE

Table 7. Handling confirmed or suspected prion-infected patients in preoperative areas

Steps in Procedure	Comments
1. Scheduling staff or OR front desk staff will notify the CN IV for Neurosurgery.	
2. CN IV will notify: <ul style="list-style-type: none"> • Pre-op (31648) • PACU (31292) • OR Charge Nurse (31580) • SPD (31084) and/or • Instrument Coordinator/Lead Tech • OR support supervisor • Infection Control (34343) • Pathology (719-9162) • OEH&S (61300) 	Notify all personnel with as much warning as possible. Verify that all equipment (instruments, cleaning equipment) is available and ready.
3. The circulating nurse will obtain the CJD cart containing supplies from the central storage area.	Contents include: <ul style="list-style-type: none"> • a copy of this policy, • yellow chemical waste bags, • blue high-risk gloves, • disposable sterile gowns, • face shields, • shoe covers, • disposable BP cuffs (both pediatric and adult) • 1N (normal) sodium hydroxide solution • 2N (normal) sodium hydroxide solution • isolation sign • tags and notification forms for OEH&S.
4. The circulating nurse will place the appropriate signage on OR doors.	
5. OR hampers and kick buckets will be lined with yellow chemical hazardous waste bags and remove all unessential equipment from the room.	Extra sharps containers can be found in the sterile supply room (SSR).
6. The circulating nurse will determine what unit the patient will be sent to post-operatively.	

2. ANESTHESIA PROCEDURE

Table 8. Handling confirmed or suspected prion-infected patients by anesthesia

Steps in Procedure	Comments
1. All anesthesia caregivers must wear Personal Protective Equipment: a. Long sleeve fluid-resistant gown 3. Eye, nose and mouth protection (e.g., mask with face shield) 4. Double gloves	Intraoperative anesthesia care will be provided by an Attending only; residents or CRNA's will not be involved.
2. All airway equipment that comes in contact with the patient's airway shall be disposed of. a. Disposable laryngeal mask airways (LMA) must be used b. Disposable laryngoscope blades and handles covered with a protective jacket must be used. c. Difficult airway equipment: In the case of a difficult airway where traditional laryngoscopy / LMA are not successful, disposable difficult airway equipment must be used, i.e., Retrograde Wire Intubation. d. All equipment that comes in contact with the patient's airway must be immediately placed into the yellow chemical hazardous waste bags for pickup by OEH&S. for incineration.	Rigid Scopes (Wu, Bullard, Upsher) and endoscopic equipment should NOT be used as incineration is the recommended treatment for critical and semi-critical instruments used in patients with known or suspected CJD. Equipment is not to contact any other surface.
3. Non-airway Equipment: Shall be processed following routine anesthesia procedure.	
4. Anesthesia Machine a. A HEPA grade filter must be attached directly to the endotracheal tube. b. The gas sampling line leading to the respired gas analyzer must be connected only to the sampling port on the circuit side of the HEPA filter. c. The machine surface must be cleaned as per routine anesthesia procedure	
5. Disposal of anesthesia equipment that contacts patient's blood or secretions a. Non-Sharps: Place in double yellow chemical hazardous waste bags for disposal. b. Sharps: Place a dedicated sharps box next to the patient and dispose immediately following use. At the end of case, close and lock this sharps box and place it in a yellow chemical hazardous waste bag.	
6. All yellow chemical hazardous waste bags shall be tagged with the words "CJD Precautions." a. Transport all yellow chemical hazardous waste bags to the OR staging area to be placed in the yellow barrel labeled CJD waste only for OEH&S pick up.	

3. INTRA-OPERATIVE PROCEDURE

Table 9. Handling confirmed or suspected prion-infected patients in operative areas

Steps in Procedure	Comments
Personal Protective Equipment (PPE) will include: <ul style="list-style-type: none"> • long sleeve disposable gowns • eye, nose and mouth protection • gloves 	
The scrub team will wear <ul style="list-style-type: none"> • head protection, • fluid-impervious sterile gowns • double gloves, • face shields • shoe covers. 	
2. Instrumentation <ol style="list-style-type: none"> a. For surgical set-up (see Preference Card) b. Obtain the dedicated CJD instrument set which includes disposable items c. Flash sterilization is contraindicated. For emergent instrument issues, page Instrument Coordinator: 719-1891. d. Empty entire 500 ml bottle of 1N sodium hydroxide into suction canister before any procedures. 	
3. Handling of Specimens: <ol style="list-style-type: none"> a. The circulating RN will invert the specimen bag to cover his/her hand while the scrub nurse is passing off the specimen. b. The circulator will invert the bag over the cup once the specimen is obtained. b. All specimens will be double-bagged and labeled with a sticker stating "CJD Precautions." c. The circulating RN will notify Pathology (719-9162) to pick up the specimen in the OR at the time of surgery and that CJD precautions are required. Specimens will be picked up immediately and not stored in the OR Pathology refrigerator. Specimens may be stored briefly in the surgical pathology refrigerator prior to final transport to the lab. 	

4. POST-OPERATIVE PROCEDURE

Table 10. Handling confirmed or suspected prion-infected patients in postoperative areas

Steps in Procedure	Comments
1. At the conclusion of the case, the circulator will notify PACU that the patient is ready for transport and that standard precautions are required for caring for patients with CJD.	
2. All linen used in the Operating Room including bed sheets, bath blankets and towels and any disposable materials exposed to any patient fluids must be placed in the yellow chemical waste bags. These bags must be placed in the yellow CJD waste tub in the specified area for CJD waste for pick up by OEH&S for incineration.	All linen used in the Operating Room including bed sheets, bath blankets and towels and any disposable materials that has not had any contact with the patient's fluids can be treated as routine soiled linen and placed in the routine soiled linen bags for routine processing.

XII. Surgical or Procedural Instrument Handling Decision-Making

Because instruments used in neurosurgical and invasive procedures in patients with confirmed or suspected human prion disease have been implicated in the transmission of prion disease in both animal studies and human cases, and because there is theoretical risk from other surgical procedures, critical decision-making about whether to destroy, disinfect and quarantine, or disinfect and reprocess instruments must be determined based upon the risk of transmission of human prion disease to patients on whom those instruments would subsequently be used. Some equipment, such as fiberoptic endoscopes and delicate ophthalmic equipment, cannot be adequately disinfected after use on a patient with confirmed or suspected human prion disease without destroying the equipment.

The division of clinical procedures into risk categories for iatrogenic transmission of prion disease is dependent on at least three factors:

- a. The type of prion disease (i.e., CJD vs. vCJD)
- b. Risk category of tissue or fluid (see Table 1)
- c. Material of equipment being used (prions adhere more readily to stainless steel and glass)

Invasive procedures on high and medium infectivity in patients with confirmed or suspected prion disease may be clinically indicated. Instruments that contact such tissues (see Table 11) must be treated as indicated. Hence, the risk of prion disease in the clinical diagnosis must be discerned prior to the procedure.

Patients with *Definite* (pathology-proven) or *Probable* (determined by Neurology service) prion disease:

Instruments must be destroyed by incineration if they contact HIGH, MEDIUM or LOW risk tissues, regardless of the instrument used.

Patients with *Possible* prion disease (patients in which prion disease is in the differential diagnosis, as determined by the Neurology Service):

Instruments must be destroyed by incineration if they contact HIGH risk tissues (see Table 11).

Instruments may be cleaned, then quarantined pending final diagnosis if they contact *MEDIUM* or *LOW* risk tissue, and can be reprocessed using routine procedures for *NO* risk tissue (see Table 12).

Patients with Asymptomatic Genetic or Iatrogenic prion disease:

Instruments must be destroyed by incineration if they contact HIGH, MEDIUM or LOW risk tissues, regardless of the instrument used.

Table 11. Directions for handling equipment used for invasive or surgical procedures in patients with confirmed or suspected prion disease.

Tissue Infectivity	Status of Patient			
	Definite/Probable	Possible	Asymptomatic	
			Genetic risk	Iatrogenic risk
High, such as:	Destroy by incineration	Destroy by incineration	Destroy by incineration	Destroy by incineration
Brain				
Spinal cord				
Invasive eye				
Lymphoreticular, in vCJD				
Medium, Low, such as:	Destroy by incineration	Clean* & quarantine pending final diagnosis ^f	Destroy by incineration	Destroy by incineration
Noninvasive eye**				
Olfactory epithelium				
Lymphoreticular in non-vCJD				
None, such as	Routine reprocessing	Routine reprocessing	Routine reprocessing	Routine reprocessing
Urine				
Sweat				
Intact skin				

*See below for cleaning instructions pending final diagnosis

**Many specific risk groups for the iatrogenic transmission of vCJD are not known. However, *blood transfusion* from a patient with vCJD is a known risk. Individual patients who have been potentially exposed to vCJD via *surgical instruments* used on a patient who went on to develop vCJD may be at higher risk than from surgical equipment exposed to other forms of prion disease, such as sCJD or gCJD. The level of prions in the *lymphoreticular system* in patients with vCJD is much higher than in patients with other forms of prion disease (i.e., sCJD or gCJD). Therefore, contact with these tissues in a vCJD patient theoretically has a higher risk of transmission. An epidemiological investigation and risk assessment should be undertaken in the case of exposure to potentially contaminated instruments or tissues.

^f As determined by the Neurology service.

Table 12. Cleaning of instruments to be quarantined:

Steps in Procedure	Comments
1. Instruments must be decontaminated by hand, with the worker wearing full protective gear.	Full protective gear includes: a. Long sleeve fluid repellent gown b. Face protection to include eyes, nose and mouth protection (e.g., mask with face shield). c. Heavy duty, water repellent work gloves.
2. All brushing to remove organic material must be done with the instrument(s) submerged in a sink of use-dilution enzymatic cleaner.	Keeping items below the surface of the solution will minimize spray from the cleaning equipment.
3. Once visible decontamination is completed, the instrument(s) will be stored in SPD, and quarantined.	
4. Allow solution in sink to flow down the drain.	Dilution is the solution: prions disposed of via sanitary sewerage with copious amounts of solution are thought to be diluted to a negligible infectious dose.
5. Flood sink and all surrounding areas with 1N Sodium Hydroxide, and allow to remain 1 hour	
6. Rinse sink and all treated areas thoroughly with water.	

XIII. TERMINAL CLEANING OF THE OPERATING ROOM

Table 13. Cleaning surgical areas

Steps in Procedure	Comments
<p>1. For cleaning the room, OR PCA's/PSA's will wear:</p> <ul style="list-style-type: none"> • Moisture impervious gowns • Mask with face shield • High-risk gloves • Shoe/leg covers if shoe contamination is anticipated. 	<p>Personnel need to be trained and observe safety guidelines when working with 1N sodium hydroxide. With the recommended precautions, it can be used safely. 1N sodium hydroxide is caustic (basic) and has a pH > 14. It causes skin burns and its effect is delayed by as much as several hours; it is readily rinsed off by water. It must not come in contact with strong acids, flammable liquids and organic halogens. Consult the 1N sodium hydroxide MSDS that must be readily available.</p>
<p>2. Decontaminate all room surfaces that have come into contact with the patient's blood and body fluids with 1N sodium hydroxide solution, including:</p> <ul style="list-style-type: none"> • OR bed, • Instrument tables, • Floor at head of bed <p>Wet surfaces and allow to sit undisturbed one hour. After one hour, rinse area thoroughly with water then clean the floor and surrounding area according to standard procedure.</p> <p>Decontaminate the remainder of the room according to standard procedure.</p> <p>To clean floor, carefully pour small amounts of 1N sodium hydroxide directly onto floor at floor level, taking care to not splash.</p>	<p>The 1N sodium hydroxide should be poured onto rags to damp wipe. To prevent creating breathing hazard, do not use spray bottles.</p> <p>Do not use mop bucket.</p> <p>Empty sodium hydroxide bottles can be disposed of with cleaning equipment</p>
<p>3. Suction all liquid wastes into one container that has been previously filled with 2N sodium hydroxide. Double bag the suction container into yellow hazardous waste bags and place into receptacle provided for liquid waste.</p> <p>4. Complete hazardous waste tag for liquid waste and affix to yellow bag.</p> <p>5. Double bag all cleaning equipment, e.g., rags and mops heads that have come in contact with 1N sodium hydroxide into another yellow hazardous waste bag.</p> <p>6. Complete Hazardous Waste tag for debris.</p>	<p>Note that 2N sodium hydroxide is used to deactivate potential prions in liquid wastes (including fluids from the field [i.e., suction fluids]), as these fluids will dilute the 2N sodium hydroxide to approximately 1N sodium hydroxide.</p> <p>Example of completed liquid hazardous waste tag is enclosed.</p> <p>Do not mix liquid waste with debris.</p>
<p>Place sharps containers in double-bagged yellow</p>	<p>Do not confuse Environmental Health and</p>

Steps in Procedure	Comments
hazardous chemical waste bag and place in the yellow CJD waste barrel in the specified area. Transport all yellow hazardous waste bags to the staging area L401 for pickup by Environmental Health & Safety (EH&S) for incineration.	Safety (EH&S) with Environmental Services (EVS).
The scrub nurse will assure that the fluid container will be double-bagged in a plastic yellow chemical waste bag and delivered to the Decontamination Area.	
The circulating nurse will fill out "EH&S Hazardous Chemical Waste Removal Form" and affix it to one of the yellow bags, and will fax the completed form to 476-0581.	Each bag must have a completed tag but only one form needs to be filled out for all yellow-bagged items. OEH&S will pick up waste M-F 8-5; or the next business day. It is safe to store the properly packaged materials in a secure area until pick-up.
At the completion of the procedure, return the CJD cart to Central Storage for inventory and restocking.	
The circulator will document in the intraoperative nursing form that the CJD protocol was followed.	

XIV. PRECAUTIONS FOR HANDLING THE DECEASED PATIENT

On the death of a patient with confirmed or suspected prion disease, the removal of the body from the unit will be carried out using normal infection control measures. It is recommended that the deceased patient be placed in a sealed body bag prior to moving. Where the skull is open or there is CSF leakage, and where sutures do not completely control this leaking, the bag will be lined with materials to absorb any fluid, and the body should be moved in a sealed body bag. Infection Control guidelines for autopsies are contained in the UCSF Department of Pathology CJD Biosafety Precautions.

XV. OCCUPATIONAL EXPOSURE

There is no evidence of occupational transmission of prion disease to healthcare workers.

A. Post-exposure Management

1. **DO NOT USE SODIUM HYDROXIDE OR BLEACH IN EYES OR MOUTH, OR ON ANY OTHER MUCOUS MEMBRANE.** For a splash to the eye or mucous membrane exposure, rinse well with saline or tap water.
2. If an exposure has occurred, i.e., contamination of skin with high, medium or low-risk body fluids or tissues (see Table 1), needlesticks or lacerations, wash affected area with 1N sodium hydroxide solution (2-3 minutes), rinse well afterwards with soap and water to neutralize the base. *Sodium hydroxide is caustic but relatively slow-acting at room temperature and can be removed from skin or clothing by thorough rinsing with water.*
3. Cover wound with waterproof dressing to prevent secondary contamination.
4. Call the Needlestick Hotline @719-3898 to report exposure.

Policy reviewed by:	Date Approved:
Infection Control Committee	10/05
Quality Improvement Executive Committee	(pending 11/05)

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