

Precautions for Handling of Specimens with Suspected or Confirmed Prion Disease
Department of Clinical Laboratories
The Ohio State University Wexner Medical Center

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Approval and Acknowledgements
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1. POLICY

- 1.1. Additional specimen handling precautions must be taken when processing and disposing of a specimen from a patient known or suspected to have a prion disease such as Creutzfeldt-Jakob disease (CJD), Gerstmann-Straussler-Scheinker syndrome (GSS), Fatal Familial Insomnia (FFI), Kuru, or other transmissible spongiform encephalopathies (TSE).

Risk of Infection	Tissue
HIGH	Brain (including dura matter), spinal cord, eye (e.g. corneas), pituitary tissues
LOW	CSF, liver, lymph node, kidney, lung, spleen, olfactory epithelium, placenta
NONE	Peripheral nerve, intestine, bone marrow, whole blood and blood products, thyroid gland, adrenal gland, heart, skeletal muscle, adipose tissue, gingival tissue, prostate, testis, tears, nasal mucus, saliva, urine, feces, semen, sweat, serous exudate and milk

- 1.2. Transmission of prion disease is by the blood and body fluid exposure route, through contact of the infectious agent through non-intact skin or mucous membranes.
- 1.3. Because prions are resistant to inactivation by normal disinfection processes, special precautions should be taken to assure that work surfaces are adequately disinfected with full strength bleach for one hour and specimens and processing material waste are packaged for incineration upon final disposition.
- 1.4. **Since the order for CJD testing may be placed as an add on subsequent to a specimen processing spill, ALL CSF SPILLS MUST BE CLEANED UP USING THE CJD PROTOCOL. BLEACH MUST BE USED TO DISINFECT ALL SURFACES WHEN A CSF FLUID IS SPILLED.**

2. PURPOSE OF DOCUMENT

- 2.1. The purpose of this document is to inform staff about special precautions to be taken during and after handling specimens that could contain prion particles. Although there have been no documented laboratory-acquired prion infections, the primary hazard is from accidental ingestion or inoculation.

3. SCOPE OF DOCUMENT

- 3.1. This policy applies to all laboratory staff who handle specimens.

4. RESPONSIBILITY

- 4.1. The Medical Directors of the Clinical Laboratories are responsible for establishing the Laboratory Safety policy. Laboratory Compliance is responsible for maintaining the policy and ensuring at least annual review.

5. PROCESS

5.1. Notification:

- 5.1.1. Notification to lab staff will be identified by the ordering of the send out test YCJD, a confirmatory test for prion disease.
- 5.1.2. Two reports will be generated from the YCJD code :
- 5.1.2.1. Pending report: generated by LIS to generate a summary of all pending CJD orders received. The purpose of this report is to alert laboratory staff that any CSF/CNS specimens must be considered potentially infectious and extra steps to decontaminate equipment where spills and splashing occur are required until a final report is received. In addition, this allows for communication to staff caring for the patient that all future CSF/CNS specimens are required to come to the laboratory labeled as potential CJD cases.
- 5.1.2.2. Resulted report: upon receipt of the send out results, a query will generate a final report stating "See IHIS for scanned report". If the results are positive for prion disease, lab staff (and other departments as needed) will be informed that extra decontamination steps must be taken for any future CSF/CNS specimens received from this patient.

Precautions for Handling of Specimens with Suspected or Confirmed Prion Disease
Department of Clinical Laboratories
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- 5.1.3. When this test is ordered, the send out tech will identify the CSF or tissue specimens with a pink label stating "suspected prion disease". These stickers are available in CPA, Critical Care, Micro Processing, and Special Functions Laboratories.
- 5.1.4. Notification may also come by noticing a note on the requisition regarding the diagnosis, through a phone call from Infection Control, or a physician. Whoever receives this information should notify their supervisor and begin the notification process.

5.2. Specimen Handling:

- 5.2.1. Procedures involving manipulation of tissue from known or suspected CJD cases must be handled under BSL-2 conditions inside a bio-safety cabinet observing universal precautions.
- 5.2.2. Do not perform frozen section testing.
- 5.2.3. If possible, isolate the specimen to avoid high traffic areas.
- 5.2.4. Place a disposable chux pad on the work surface inside the bio safety cabinet to contain spillage/splashing and remove any unnecessary items from the processing/testing area.
- 5.2.5. Disposable supplies or equipment must be used whenever possible to reduce contamination and facilitate cleanup and disinfection.
- 5.2.6. Procedures involving aerosolization such as centrifugation, sonication and/or laser dissection pose the greatest risk to personnel.
- 5.2.7. Reduce the risk of splashes or aerosols by capping cytocentrifuge funnels while in use.
- 5.2.8. Microtome blades and knives used for cutting tissue must be cleaned with an instrument that does not put the employee's hands or fingers near or in contact with the blade.

5.3. Specimen storage:

- 5.3.1. After processing and/or testing, assure the specimen is tightly capped. Specimens must be double-bagged using biohazard waste bags, and the specimen bag must be labeled with the pink "suspected prion disease" sticker. The double bagged specimens should be given to Micro processing at UH or Microbiology at UHE, who will store the specimen for the usual 7 days.

5.4. Post Processing Decontamination and of the work area:

- 5.4.1. After initial processing, discard all consumable items and disinfecting materials (chux used during processing, paper towels, gauze) into a double bagged biohazard trash bag that will be taken to Micro processing at UH or Microbiology at UHE and placed into a biohazard bag lined corrugated incineration box.
- 5.4.2. Dispose of culture loops, scalpel blades, tissue grinder & other sharps in an appropriate biohazard sharps containers and then close to be removed from the area and placed inside the incineration waste box.
- 5.4.3. Disinfect work surfaces that were protected with chux (or other protective material) per normal protocol, unless high infectivity sample or unless there is an uncontained spill of a low infectivity sample.
 - 5.4.3.1. ***PROCESSING OF a high infectivity specimen (see Table above): soak hemocytometers, cytospin clips or other re-usable items in BLEACH for an hour, and then rinse with water and air dry. Wipe out the inside of the bio safety cabinet and the cytocentrifuge, if used, with BLEACH, let it stand for an hour and then rinse with water.***
 - 5.4.3.2. ***SPILLS of CSF & other low infectivity samples (see Table above) OUTSIDE of the bio safety cabinet: absorb with chux and soak the area in BLEACH for an hour, and then rinse with water and air dry. If spill occurs in the cytocentrifuge, wipe out the inside of cytocentrifuge with BLEACH, let it stand for an hour and then rinse with water.***

Precautions for Handling of Specimens with Suspected or Confirmed Prion Disease
Department of Clinical Laboratories
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5.5. Final Waste Disposal: (Incineration)

- 5.5.1. All specimens from an individual with confirmed or suspected Prion Disease/CJD, must be double bagged and placed into a biohazard bag lined corrugated box that contains the double-bagged consumable items and disinfection materials used in the handling/processing of the sample.
- 5.5.2. Tie off the outer bag by gathering and twisting the neck of the liner and hand knot (Gooseneck tie).
- 5.5.3. Close the top and bottom of the box and tape with a 2 inch wide pressure sensitive tap or equivalent.
- 5.5.4. Ensure box is labeled “INCINERATE ONLY” with the EVS-provided labels.
- Both laboratory areas (UHE and UH) have the corrugated incineration boxes, large biohazard bag and labels stored in their respective areas.
 - For additional supplies (corrugated boxes and incineration labels), UHE will need to call EVS at UHE and UH will need to call EVS at Main.
- 5.5.5. The box **should not be marked Prion Waste and/or CJD waste**, if the box is marked as “Prion Waste” or “CJD”, Stericycle will NOT remove from OSUWMC.
- 5.5.6. Stericycle will not accept the waste if labeled Prion Waste and if not in the labeled incineration boxes.
- 5.5.7. Final disposal of the waste will be via incineration only at a designated Stericycle incineration facility.

5.6. Special Considerations for Surgical Pathology

- 5.6.1. Single-use protective clothing should include: gloves, disposable laboratory coat/protective apron and sleeves, face shield.
- 5.6.1.1. For tissues, secretions, or excretions with low or no detectable infectivity (see chart above), no special requirements beyond Standard Precautions are required for the handling of body fluids or body-fluid contaminated linen, equipment or environments.
- 5.6.2. **No frozen sections will be performed on tissue confirmed or suspected of prion disease. Any frozen section form completed with a CJD sticker and pre-op diagnosis of “potentially contaminated with prions (CJD)” should induce a hard stop.**
- 5.6.3. To avoid cross contamination with subsequent cases, the following precautions should be taken for tissue types that pose a low or high risk of infectivity:
- 5.6.3.1. Cut surgical specimens on a clean chuck/diaper placed on top of a clean cutting board, but preferably, within the lid of the container as to not contaminate any laboratory surfaces.
- 5.6.3.2. Use disposable forceps and scalpel/razor blade.
- 5.6.3.3. Change gloves when case is complete.
- 5.6.4. The following will occur in the operating room for suspect CJD cases:
- 5.6.4.1. When a case potentially contaminated with prions (CJD) is scheduled in the OR, the circulating nurse will call Pathology to alert them of the procedure.
- 5.6.4.2. When specimens are collected, they are placed in appropriately labeled containers including “potentially contaminated with prions (CJD)” written after the tissue description, as well as, have a bright sticker labeled “potentially contaminated with prions (CJD)” applied elsewhere on the container.
- 5.6.4.3. The specimen order is entered into IHIS and “potentially contaminated with prions (CJD)” is typed into the description box next to the tissue description. A bright sticker labeled “potentially contaminated with prions (CJD)” is also applied to the requisition.

Precautions for Handling of Specimens with Suspected or Confirmed Prion Disease
Department of Clinical Laboratories
The Ohio State University Wexner Medical Center

- 5.6.4.4. When manually filling out a frozen section form, the pre-op diagnosis should state “potentially contaminated with prions (CJD)” and a bright sticker labeled the same will be applied to the frozen section requisition.
- 5.6.5. Discussion with the neuropathologist on-service, Director of Neuropathology) or Director of Anatomic Pathology (if the neuropathologist is not available) should occur before deciding whether the specimen should be partly processed or whether the entire specimen should be sent to the CJD Surveillance National Prion Disease Pathology Surveillance Center in Cleveland, Ohio.
- 5.6.5.1. If any part of the specimen is to be sent to the surveillance center, then the testing form should be printed from Q-Pulse, thoroughly completed and the specimen should be packaged and shipped as per instructions on www.cjdsurveillance.com.
- 5.6.5.2. If all tissue is not to be sent to the surveillance center, snap freeze 30% of the specimen and store in the -80 freezer until results are received from the surveillance center.
- 5.6.5.3. Write the measurements and description of the entire specimen on a frozen section form and store with the frozen tissue. Indicate on the form how much tissue was sent to the surveillance center.
- 5.6.5.4. To snap-freeze, do not dip the tissue into the liquid nitrogen container. Under the hood, ladle the liquid nitrogen over the mold filled with the OCT and tissue over a metal receptacle.
- 5.6.6. A result from the surveillance center must be received before CJD-suspected tissue will be processed at OSU Wexner Medical Center.
- 5.6.6.1. If negative results are received from the surveillance center, submit the tissue for processing
- 5.6.6.2. If positive results are received from the surveillance center, the tissue will be disposed as per the appropriate waste disposal protocol:
- Double-bag the chuck/diaper, non-sharp disposable instruments and PPE. Tie off the outer bag by gathering and twisting the neck of the liner and hand knot (gooseneck tie).
 - Label the bag “suspected CJD waste” and immediately transport the bag to the Morgue for proper disposal.
 - Once in the morgue, the double-bagged CJD waste bag will be placed into a biohazard bag-lined corrugated box and be sent for incineration
 - Discard the disposable scalpel/razor in the sharps container.
 - Flood and/or soak any non-disposable instruments and surfaces with household bleach (6.15%) for one hour, followed by a water rinse.
- 5.7. **Autopsy Requests on Patients with Confirmed/Suspected Prion Disease**
- 5.7.1. The Division of Autopsy Services does NOT perform autopsies on individuals with confirmed or suspected prion diseases, such as CJD.
- 5.7.1.1. An adequate review of the clinical history is critical in the determination of potential cases of prion disease with particular attention directed toward the following signs/symptoms:
- Rapidly progressive dementia of less than three years duration (typically less than one year).
 - Dementia with seizures, particularly myoclonic seizures.
 - Dementia associated with cerebellar or lower motor neuron signs.
- 5.7.2. If an autopsy is requested on a patient with confirmed or suspected prion disease, the next-of-kin (NOK) and the medical team are directed to the National Prion Disease Pathology Surveillance Center (NPDPSC), which operates out of Case Western Reserve University in Cleveland, Ohio. The disposition of the decedent remains at the preference of the NOK.

Precautions for Handling of Specimens with Suspected or Confirmed Prion Disease
Department of Clinical Laboratories
The Ohio State University Wexner Medical Center

- 5.7.2.1. OSUWMC staff may aid in printing documents, but may not facilitate consent for NPDPC. All transportation arrangements, clinical information exchange, and consents must be orchestrated by NPDPC and the NOK in conjunction with the OSUWMC neurologist.

National Prion Disease Pathology Surveillance Center
Institute of Pathology
Case Western Reserve University
2085 Adelbert Road, Room 418
Cleveland, Ohio 44106-4907
Tel: 216-368-0587
Email: cjdsurv@case.edu
<http://www.cjdsurveillance.com/>

6. REFERENCES

- 6.1. CDC recommendations for infection control in Prion Disease cases:
<https://www.cdc.gov/prions/cjd/infection-control.html>
- 6.2. [WHO Infection Control Guidelines for Transmissible Spongiform Encephalopathies](#): Report of a WHO consultation, Geneva, Switzerland, 23-26 March 1999
- 6.3. CJD Surveillance website, National Prion Disease, Pathology Surveillance Center, Case Western Reserve University
- 6.4. Stericycle, Inc Policy Manual; Policy number SH-024, effective date 3/18/2015

7. RELATED DOCUMENTS

- 7.1. Refer to Q Pulse System or Document Detail Report for related Laboratory Policies, Procedures, and Master Forms