

## Laboratory Guidelines for Creutzfeldt-Jakob Disease (CJD)

Special precautions are required when collecting, handling and transporting samples from a suspected CJD or TSE patient. The requirements for submitting these types of specimens are outlined below.

**Background:** Transmissible Spongiform Encephalopathy (TSE) disease refers to a group of diseases, inherited or transmissible that may occur in animals and humans that share common characteristics related to the deterioration of the central nervous system. There is no known treatment, and the outcome is uniformly fatal.

This group of diseases includes: Creutzfeldt-Jakob Disease (CJD), Gerstmann-Straussler-Scheinker Syndrome (GSS), Fatal Familial Insomnia (FFI), and Kuru. The prion proteins associated with these diseases are hardy, remain intact for years in a dried state, and resist all routine sterilization and disinfection procedures commonly used in healthcare facilities.

Based on epidemiological evidence, the World Health Organization (WHO) recommends that:

Blood and /or its components require no special precautions for handling in clinical laboratories. Standard precautions apply.

With the exception of Cerebrospinal Fluid (**CSF**), other body fluids, secretions and excretions need no special handling.

Tissue with high concentrations of prion proteins require special precautions. (e.g. cornea, brain, spinal cord). Note: although infectivity has been detected in CSF, it is considered a lower risk than brain tissue.

### **CLINICIAN PROCEDURE FOR CSF COLLECTION for Protein 14-3-3 Testing:**

**PRIOR** to collecting the CSF specimen, the attending physician and/or neurologist **MUST** call ) the Central Zone Pathologist on call (403-343-4422) who will contact the UAH or CLS Microbiologist on call (MOC) to discuss testing of samples from high risk patients and to seek approval of 14-3-3 protein testing where deemed appropriate.

The Pathologist on Call will contact the appropriate MOC, depending on the location of the patient. Samples from patient in Central Zone will be referred to Edmonton (UAH) with the exception of Drumheller, Hanna and Three Hills. Those three sites will refer to Calgary (CLS).

The contact number for the UAH Microbiologist On Call is 780-407-7121.

The contact number for the CLS Microbiologist On Call is 403-770-3757.

In general, the patient must have shown rapid, progressive dementia, with at least one of the following clinical findings:

- Patient has had an enhanced MRI with changes.
- Patient has EEG changes consistent with possible CJD/TSE.
- Patient shows signs of myoclonus.

**AFTER** obtaining approval, the Central Zone Pathologist On Call will contact the attending physician and/or neurologist to confirm approval of testing, **ensure specimen is labeled “CJD precaution” and requisition is clearly labeled to identify the specimen as “CJD/Prion Agent”**.

**LABORATORY STAFF PROCEDURE:**

- Central Zone Pathologist on Call will contact appropriate Central Zone lab site if specimen is approved for referral for CJD or 14-3-3 protein testing.
- Central Zone Pathologist on Call must also inform Red Deer Hematology, Microbiology and Cytology that a specimen for CJD is expected and what tests are being approved for each area.
- Red Deer Hematology will perform a cell count only, using a disposable counting chamber loaded within a Class II biological safety hood. Refer to HEM.2110 Appendix A.
- Cytology and Microbiology will only proceed with testing when advised by the pathologist on call. All testing will be performed manually using disposable material and containers.
- Chemistry will NOT perform any testing.
- All material used will be disposed of following CJD precautions.
- **Ensure “CJD precaution” is clearly indicated on all specimens and requisition is labeled to identify the specimen as “CJD/Prion Agent”.**

**PRION DISEASE AUTOPSY REQUESTS:**

CJD Autopsy/Anatomical Pathology cases will be dealt with ONLY at Calgary Laboratory Services (CLS) and University of Alberta Hospital (UAH). Attending physicians are to contact Central Zone Pathologists on Call for autopsy request. Central Zone Pathologist will contact CLS Neuropathologist on-call to inform them of pending CJD Autopsy and to get their approval for acceptance of the case. Central Zone refers any CJD/suspect CJD cases to the Foothills Medical Centre Laboratory (CLS).

**RELATED DOCUMENTS:**

- Prion Disease (CSF & Tissue) Communication & Testing Process for the Laboratory
- CJD CSF & Tissue Specimen Transport and Receipt
- Clinical Laboratory Testing Areas – CJD CSF & Tissue Safe Work Practices
- CJD Decontamination & Disposal Safe Work Practices
- Autopsy / Anatomic Pathology – CJD Tissue Safe Work Practices