

**Creutzfeld-Jakob Disease:
Why Should Laboratory Workers Protect Themselves? – March 1999**

Creutzfeld-Jakob Disease (CJD) is one of a group of related diseases known as Transmissible Spongiform Encephalopathies (TSE). CJD is a rare degenerative disease which attacks the brain and other tissues of the central nervous system. There is no effective treatment and it is always fatal. Once symptoms appear the disease progresses rapidly. Symptoms include dementia, progressive motor dysfunction and personality changes. Death usually occurs within one year of the onset of symptoms.

There are three forms of CJD – sporadic, familial, and infectious/iatrogenic. Sporadic CJD accounts for 90% of cases, occurs at random in the population and usually affects patients who are in middle age (60-65 years). Familial CJD is thought to be linked to gene mutation and is responsible for 5 – 10 % of cases. CJD related to an infectious agent is thought to be responsible for less than 1% of cases and is the result of transmission during medical treatment.

The causative agent is thought to be an abnormal form of a protein particle known as a prion. It is smaller than a virus and contains no nucleic acid. It is a very hardy pathogen which can remain infectious for years. The incubation period can be as long as 30 years, making it difficult to trace the source of infection. It resists routine sterilization and disinfection techniques commonly used in health care facilities. It is resistant to heat, formalin, glutaraldehyde, radiation, freezing, organic detergents and ethylene oxide.

Iatrogenic transmission occurs during person-to-person transplant of corneas or dura matter grafts, use of contaminated neurosurgical instruments or injection of human cadaveric growth hormone or human pituitary gonadotropin from donors with CJD.

There is no strong evidence to suggest CJD can be transmitted from patients to health care workers although isolated cases have been reported. It must therefore be considered a possibility.

What precautions are required of laboratory personnel when handling specimens from CJD cases?

The brain, spinal cord or other tissues from the central nervous system, eyes, pituitary, cerebrospinal fluid, tonsil tissue, lymphatic tissue, lymph nodes and thymus from known or suspected cases of CJD present a high level of risk of infectivity.

Blood, bone, marrow, placenta, nasal mucus, semen, sputum, urine, stool, tears, saliva, sweat and other external secretions are estimated to present low or no risk of infectivity.

Universal Body Substance Precautions should be followed when handling any specimen from a patient who is diagnosed or suspected of having CJD. In addition, there are some laboratory procedures such as autopsy or histological examination of brain or other tissue from the central nervous system that require additional safety measures. Since the causative agent is difficult to destroy, decontamination and waste disposal become a concern.

Each laboratory must develop its own policies and procedures that describe in detail, the steps to follow when confronted with CJD. Staff must be trained and be able to demonstrate knowledge of the procedures.

In addition to Universal Body Substance Precautions, procedures should at least include the following:

- Wear personal protective clothing, ie: gown, double gloves, face mask, goggles
- Use disposable linens, supplies and equipment where possible and incinerate after use
- Decontaminate soiled work surfaces with 2N NaOH of 60 minutes or 2.5% bleach for 60 minutes
- Dispose of all waste materials by incineration, ie: specimens, wash liquids
- Re-usable instruments should be dedicated solely for use with CJD cases. Decontaminate by steam autoclaving at 134 C for 60 minutes or soaking in 2N NaOH for 60 minutes
- Tissues for histological examination should be fixed in formalin followed by fixation in formic acid
- Take care to avoid puncture wounds or the creation of aerosols
- Use manual saws; no power tools.

Procedures for histology and autopsy examinations require more detail than can be described in this article. For further reading, refer to the references.

References:

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