POLICY & PROCEDURE NUMBER: 4.30

DEPARTMENT: *Infection Control* **SUBJECT:** *Management of Creutzfeldt-Jakob Disease (CJD)*

APPROVAL DATE: April 2004 **REVISION DATE:** March 2016; Nov. 2022
REVIEW DATE: March 2017; Nov 2018; Dec 2019; Dec. 2020

MANAGEMENT OF CREUTZFELDT-JAKOB DISEASE (CJD)

CJD was first identified in the 1920s. It is one of a group of diseases known as transmissible spongiform encephalopathies (TSE). TSEs can occur in humans or animals and cause deterioration of the central nervous system. CJD is a form of TSE that occurs only in humans.

CJD occurs at a worldwide rate of between 0.5 and 1 case per million population per year. The distribution of cases is evenly divided between men and women and the age of onset is between 50 and 75 years of age.

CJD differs from variant CJD (vCJD) which has been linked to Mad Cow Disease in the United Kingdom and Europe. Variant CJD has only been seen twice in Canada (one case each in 2002, and 2011.)

The agent that causes CJD is thought to be an unconventional agent known as a prion protein. The prion contains only protein that replicates by converting normal cellular protein to abnormal protein.

CJD leads to deterioration of the central nervous system. The initial stages of the disease are subtle and may involve symptoms of insomnia, depression, confusion, personality and behavioural changes, strange physical sensations, and problems with memory, co-ordination and sight. As the disease advances, the individual experiences a rapidly progressive dementia and in most cases, involuntary and irregular jerking movements known as myoclonus. Problems with language, sight, muscular weakness, and co-ordination worsen. In the final stage of the disease, the individual loses all mental and physical functions and may lapse into a coma. The duration of the illness is generally short but may vary from several weeks to several years. CJD is always fatal.

Diagnosis of CJD is extremely difficult. At the present time, the only accurate way to make a diagnosis of CJD is by autopsy. An experimental test is available, but the low sensitivity and specificity of the test make it unreliable. Other brain disorders may result in a positive test. Diagnosis is made principally by clinical and neuropathological examination.

Three forms of CJD are recognized: sporadic, familial and infectious/iatrogenic. Transmission of CJD varies depending on the type of illness.

Sporadic CJD

Sporadic CJD accounts for 85-90% of all cases in the general population. It occurs at random and there is no known infectious source. There is no evidence of the disease being transmitted in the patient's family.

1. **Familial CJD**
   The familial form accounts for 5-10% of all cases and is inherited. These cases exhibit a mutation in the gene coding for the prion protein.

2. **Infectious CJD**
   This form is very rare, occurring in less than 1% of cases. Disease is the result of transmission during an invasive medical procedure (e.g. brain surgery, corneal transplants). There are three circumstances in which transmission of CJD between humans has been demonstrated: by transplantation of central nervous system tissue (dura mater grafts), by contaminated instruments used during invasive neurological or neurosurgical procedures, and by administration of human pituitary extracts. Person-to- person transmission by any other route has not been demonstrated.
   Family members who live with infectious CJD patients have no greater risk of getting CJD than the general population.

CJD is not known to be transmitted through contact with blood or body fluids other than brain tissue and possibly cerebrospinal fluid (CSF). No special precautions are required in the long-term care setting. Routine Practices for infection prevention should be used with all residents.

Long-term care staff and residents are not at any increased risk from a patient with CJD. Residents with CJD may participate in all activities within the facility.

Staff education should be done whenever a case of CJD is identified in the facility.

References:

Public Health Agency of Canada, Creutfeldt-Jakob Disease Surveillance System (CJDSS) Report; Available at: https://www.canada.ca/en/public-health/services/surveillance/blood-safety-contribution-program/creutzfeldt-jakob-disease/cjd-surveillance-system.html
Ontario Health Association (2019) Appendix A Creutzfeldt-Jakob Disease, all types. Available at:
https://www.health.gov.on.ca/en/pro/programs/publichealth/oph_standards/docs/cjd_chapter.pdf

Public Health Agency of Canada Classic Creutzfeldt-Jakob Disease in Canada Quick Reference Guide 2007, Available at https://www.phac-aspc.gc.ca/nois-sinp/pdf/cjd-eng.pdf

Weinstein, R., Rutala, W., Weber, D., Creutzfeldt-Jakob: Recommendations for Disinfection and Sterilization Clinical Infectious Diseases Vol 32, Issue 9 May 2001 P 1348-1356 Available at: https://academic.oup.com/cid/article/32/9/1348/291736

Human Transmissible Spongiform Encephalopathies. January 1 1999; Vol 25-1:1-7 unable to locate

Control Guidelines: Routine Practices and Additional Precautions for Preventing the Transmission of Infection In Health Care. July 1999; Vol 25S4. Unable to locate

1998; P 12-14.