Creutzfeldt-Jakob Disease (CJD)

Site Applicability

All PHC Acute and Long-term Care Sites

Practice Level

Basic: Physicians, NPs, Nursing, Clinical Nurse Leader, Clinical Site Coordinator, Bed Placement Coordinator

Standards

Routine Practices will be used on all patients/residents with suspected or confirmed Creutzfeldt-Jakob Disease (CJD). No Additional Precautions are required.

Special handling/reprocessing of critical instruments that come into contact with body fluids and tissue potentially infectious for CJD is required:

- **High-infectivity:** brain, cerebral spinal fluid (CSF), dura mater, pituitary gland, posterior eye (optic nerve and retina), spinal cord and spinal ganglia, trigeminal ganglia.
- **Low-infectivity:** cornea, kidney, liver, lung, lymph nodes, placenta, and spleen
- **No infectivity:** all other body tissues and organs not listed above, blood and body secretions/excrement including saliva, sweat, breast milk, semen, tears, feces, and urine

Description of the Disease

Creutzfeldt-Jakob Disease is a neurodegenerative disease caused by the deposition of a unique protein called a “prion” into brain tissue. This eventually leads to dementia that may appear similar to other disorders like Alzheimer’s disease, but the progression is much more rapid. No treatment exists for CJD, and all cases are fatal. Prion are very stable and resist most decontamination methods.

Most people with classic CJD develop the disease for no apparent reason, which is called spontaneous CJD or sporadic CJD. Around 15% of cases may have familial CJD, which is the result of a genetic mutation. A small number of people have acquired CJD after being exposed to infected human tissue during a medical procedure (iatrogenic CJD; e.g., corneal transplant, brain surgery using contaminated instruments) or from eating beef with bovine spongiform encephalopathy, or mad cow disease (variant CJD).

Signs & Symptoms

CJD is marked by rapid mental deterioration, usually within a few months. Early signs and symptoms typically include:

- Personality changes
- Memory loss
- Impaired thinking
- Blurred vision or blindness
- Insomnia
- Incoordination
- Difficulty speaking
- Difficulty swallowing
- Sudden, jerky movements

As the disease progresses, mental symptoms worsen. Most people eventually fall into a coma. Heart failure, lung (respiratory) failure, pneumonia or other infections are generally the cause of death, which usually occurs within a year.

Incubation Period

For acquired CJD the incubation period can be from 15 months to possibly more than 30 years.

Period of Communicability

Central nervous system tissues and fluids are infectious throughout symptomatic disease. Level of infectivity during the incubation period is unknown.

Routes of Transmission

The risk of CJD transmission is low and requires contact with high-infectivity tissue of a CJD patient through contaminated instruments. The disease cannot be spread through coughing or sneezing, touching, or sexual contact.

Assessment and Intervention

Infection Control Precautions

- **Additional Precautions:** No Additional Precautions are required above Routine Practices.
- **Hand Hygiene:** Hands should be cleaned before and after every patient contact, as well as after touching items in the environment per Routine Practices. Using an alcohol based hand rub solution is preferred if hands are not visibly soiled.
- **Patient Placement:** Patients with CJD may be placed in any available bed/room and can safely share a room with other patients.
- **Non-Critical Equipment:** Clean and disinfect shared, non-critical equipment routinely and between different patients/residents per Routine Practices.
- **Critical Instrumentation and Supplies Used in a Surgical Procedure:**

  If a presumed or confirmed CJD patient/resident requires booking for a surgical case that may include at least one surgical procedure involving contact with high- or low-infectivity tissue, surgeon consultation with Medical Device Reprocessing Department (MDRD) should ideally take place a minimum of 10 business days in advance of an elective surgical case, or as soon as

possible for an urgent surgical case. The surgeon and MDRD will negotiate options and a plan for a CJD specific pick-list for any procedure at risk of surgical contact with CJD infected tissue in addition to consultation regarding instrumentation required for any procedure that is not at risk of surgical contact with CJD infected tissue.

For instrumentation at risk of contact with CJD infected tissue, preference will be for:

- Disposable, single use instrumentation and supplies whenever possible.
- Non-disposable instrumentation that would be deemed suitable to have destroyed post procedure due to age, condition or low demand
- Non-disposable instrumentation that is validated by the manufacturer to tolerate the precautionary CJD decontamination procedure required to complete a terminal kill of infected tissue that may contain prions.

The CJD decontamination procedure involves a high dwell time in concentrated, alkaline chemistry and can both damage instrumentation and be hazardous to MDRD staff if materials within the instrumentation reacts to the chemistry.

Post-procedure, all unused and unopened instrumentation and supplies shall be sent back to MDRD as per normal procedure for clean flow. Any possibly CJD contaminated instrumentation must be segregated from any unsterile (e.g. open, but unused) or non-CJD contaminated instrumentation (e.g. any instrumentation that was used for a procedure in a surgical site that has no risk of CJD presence). If necessary, the OR can call MDRD to send up an empty case cart to contain the unsterile and/or non-CJD contaminated instrumentation to be sent to MDRD through routine, soiled process. However, potentially CJD contaminated instrumentation must be contained in a separate case and labeled for CJD reprocessing to be maintained in quarantine until sent to MDRD for review and next steps. Any supplies opened in the OR would be discarded in the OR as per procedure.

In the event that a patient is presumed or confirmed positive for CJD, post-surgical procedure, IPAC should be alerted immediately so that MDRD can identify instrumentation used and immediately quarantine as part of an investigation process.

- **Environment:** No special/additional environmental cleaning is required. All high-touch surfaces in the patient's room must be cleaned and disinfected at least daily. Following discharge of the patient, the room should have a terminal clean carried out prior to the next patient being admitted.
- **Visitors:** Education should be provided regarding hand hygiene.
- **Patient Transport:** No added measures are required during transport.

Lab Testing

- Diagnostic testing for CJD is available for CSF. If CJD is on the differential diagnosis and CJD testing on CSF is requested, it is important to notify the laboratory so that appropriate lab biosafety protocols can be utilized.

- Post-mortem, an autopsy may be requested to confirm the diagnosis of CJD from brain tissue.

Treatment

- No treatment exists for CJD.

Transfer/Discharge Planning

- Notify the receiving facility, hospital, nursing home or community agency involved in the patient's care of their status.

Patient and Family Education

HealthLinkBC Files:

- Creutzfeldt-Jakob Disease (CJD)

Related Documents

- B-00-07-13045 – Routine Practices - Infection Control

References

Alberta Health. (2022). Alberta Public Health Disease Management Guidelines: Creutzfeldt-Jakob Disease (CJD). Available from https://open.alberta.ca/dataset/e45caa5c-00a1-4fe6-83a8-7695872978a2/resource/93c64c22-65c4-460e-b992-7d30b848a3bb/download/health-phdmg-creutzfeld-jakob-disease-2022-02.pdf

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Uttley, L., Carroll, C., Wong, R., Hilton, D. A., & Stevenson, M. (2020). Creutzfeldt-Jakob disease: a systematic review of global incidence, prevalence, infectivity, and incubation. *The Lancet. Infectious diseases*, *20*(1), e2–e10. https://doi.org/10.1016/S1473-3099(19)30615-2

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