Creutzfeldt-Jacob Disease and Other Prion Diseases:

A Challenge for the Decontamination Department

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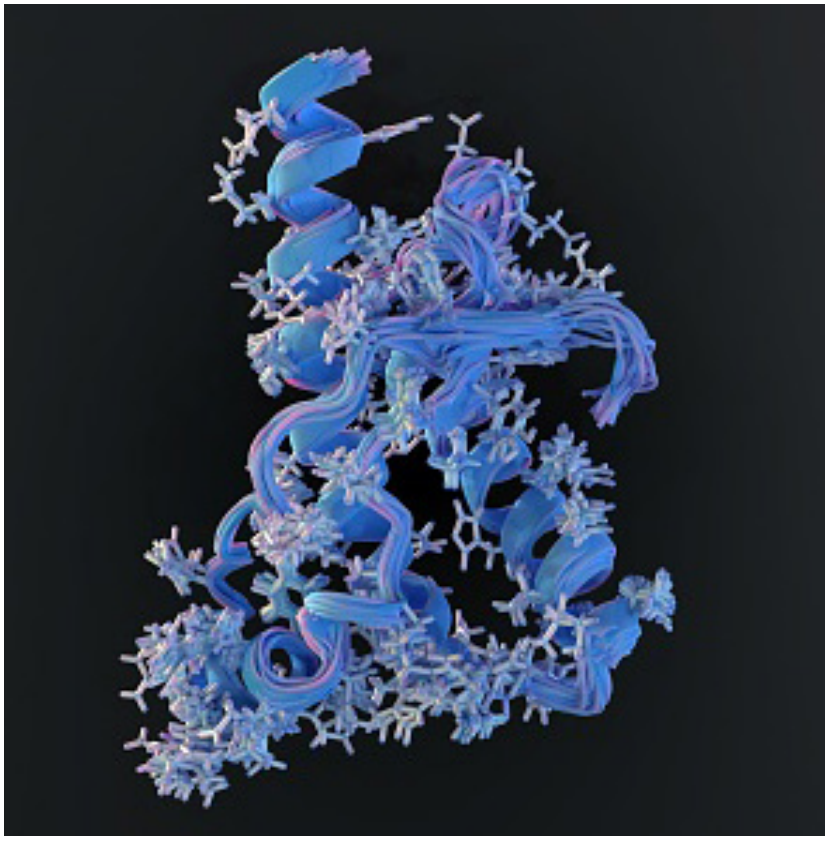
Creutzfeldt-Jacob Disease (CJD) is one of several transmissible spongiform encephalopathies (TSEs), which are characterized by the accumulation of abnormal forms of the prion protein.[1] Clinically, CJD presents as a progressive deterioration of brain function arising from the buildup of misfolded proteins within the brain, which impair normal function, ultimately causing tissue damage. Histological examination indicates the formation of microscopic vacuoles within brain tissue, making it appear sponge-like, hence the use of the term "spongiform". All known prion diseases are invariably fatal.

There are a number of mammalian TSEs that have been known for centuries, the most common being scrapie, which affects sheep. Other mammalian TSEs include chronic wasting disease (CWD) in elk and deer. More widely known is bovine spongiform encephalopathy (BSE), also known as mad cow disease, which occurs in cattle. Several human forms of TSEs exist, the most well-known being CJD. Another is kuru, identified in tribal communities in Papua New Guinea whose funerary traditions included cannibalism. Other human forms include Gerstmann-Sträussler-Scheinker syndrome and fatal familial insomnia.[1, 2]

Learning Objectives

1. **Describe routes of prion transmission relevant to sterile service departments.**
2. **Explain the limitations of conventional reprocessing on prions.**
3. **Identify sterile processing practices used to manage potential prion exposure.**
4. **Recognize limitations of sterilization technologies and how monitoring systems affect prion risk management in sterile processing.**



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The aim of this lesson is to introduce sterile services technicians to key concepts related to prion transmission risk and its implications for sterile processing.

LEARNING OBJECTIVE 1:

Describe routes of prion transmission relevant to sterile service departments.

Unlike diseases caused by bacteria or viruses, which involve replication of an organism, prion diseases progress through the propagation and accumulation of misfolded prion proteins. These misfolded proteins act like a template that induces normally folded proteins to adopt the same abnormal conformation, initiating a self-propagating chain reaction. The misfolded proteins accumulate because the body's normal functions are unable to eliminate them. The method of diagnosis relies on the development of symptoms and supporting brain scans; a definitive diagnosis is made after the disease has progressed.

Prior to 1980, prion diseases received limited attention due to their infrequent incidence, occurring at a rate of approximately one to three cases per million globally.[1] However, in the 1980s, the incidence of BSE rose dramatically in the United Kingdom (U.K.) and was subsequently found to be due to a change in cattle feed processing that allowed prions to enter the food. This would have been considered a serious but manageable veterinary incident until human cases of CJD appeared in the U.K. population. These cases occurred in young individuals and proved fatal within months. Investigation indicated a causal relationship between the ingestion of BSE-contaminated cattle meat and the development of the disease, resulting in the identification of a new variant of CJD (vCJD).

This discovery became a major health crisis, resulting in the implementation of stringent control measures and the instigation of research to understand the cause, routes of transmission, and consequences on human health. Although the highest level of concern occurred more than four decades ago, studies have shown that there could be a residual population of vCJD carriers, making ongoing preventative measures important.

CJD and other TSEs can occur in several ways, including the following:[1, 2]

- Sporadically, with few cases available to study causal relationships.
- Genetically, due to inherited mutations in the prion protein gene.
- Through oral ingestion of contaminated food, such as meat derived from TSE-infected animals. This is thought to be the pathway of the vCJD outbreak in the U.K.
- Iatrogenically as a result of
- Medical procedures involving exposure or transplantation of materials or tissues containing prions. Historically, reported sources have included transplantation of dura mater and use of human-derived therapeutic agents such as human growth hormone extracted from pituitary glands.[1,2,3]
- Improperly decontaminated reusable surgical instruments previously used on a patient with a TSE. A primary example of this involved intracranial electroencephalography electrodes contaminated with prion protein from previous use on a patient with CJD. The electrodes were subsequently used on a second patient, resulting in transmission of disease. Other laboratory studies have supported this mode of transmission.[1,3]

The infectious dose of a TSE agent is thought to be very low. Experimental studies indicate that small amounts of disease-associated prion protein may be sufficient to initiate disease, depending on the route of exposure, such as oral

Lesson:

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Quiz Answers:
1. A, 2. C, 3. B, 4. C, 5. C, 6. B, 7. C, 8. C, 9. B, 10. B, 11. A, 12. C, 13. C, 14. A, 15. C

or direct exposure to brain tissue. The risk of transmission is tissue-dependent, with brain, spinal cord, and eye tissues classified as having higher levels of prion infectivity in patients with CJD.[3]

Learning Objective 2: Explain the limitations of conventional reprocessing on prions.

National guidance must be consulted to develop operational policies and protocols for prion risk management. The World Health Organisation (WHO) issued guidance on managing risks associated with TSEs, including considerations for contaminated reusable medical devices.[3] Individual countries have implemented this guidance directly or adapted it based on national expert advice and opinion, regulatory frameworks, local circumstances, and assessed risks.[4,5]

Prion proteins are chemically stable and therefore resistant to many of the conventional processes used for disinfection and sterilization of medical devices. A number of methods have been described to remove prion contamination from surfaces or reduce infectivity.[6] Many of these methods have been developed within laboratory settings and are not practical for use in a sterile service department (SSD).

The WHO recognizes that prions are unusually resistant to the cleaning, disinfection, and sterilization processes routinely used in SSDs. As a result, standard reprocessing cycles cannot be relied upon if medical devices become contaminated. WHO guidance references enhanced decontamination strategies that have demonstrated greater effectiveness under research and controlled conditions. These approaches include extended exposure times at elevated steam-sterilization temperatures, use of strong chemical agents such as sodium hydroxide or high concentrations of sodium hypochlorite, and thorough cleaning. However, the WHO acknowledges that many of these methods raise practical challenges for SSDs, including concerns related to staff safety, handling of hazardous and corrosive chemicals, equipment compatibility, and potential damage to surgical instruments. Use of such chemicals in an SSD can create operational problems and safety concerns.[6]

Because no monitoring tool can measure prion risk reduction, SSDs must rely on process assurance rather than biological confirmation.[3, 4, 11] Routine monitoring of every cleaning, disinfection, and sterilization load, using appropriate mechanical data as well as chemical and biological indicators, helps demonstrate that process parameters have been met, supports appropriate load-release decisions, and helps ensure documentation, traceability, and compliance.

Learning Objective 3: Identify sterile processing practices used to manage potential prion exposure.

One approach to managing prion transmission risk involves single-use instruments and the quarantine and disposal of reusable instrument sets used on high-risk tissues or on patients known to have a high risk of carrying a TSE disease. This approach presents the lowest risk of transmission from one patient to another but is expensive since surgical instrument sets are quarantined whilst awaiting confirmation of disease and then destroyed by incineration if disease is confirmed. Single-use instruments are invaluable when following this approach. This approach relies on clear identification of patient risk, based on a reliable clinical diagnosis of a TSE or assessment of known risk factors, such as family history; previous therapeutic interventions; and recognized non-medical exposure routes, including ingestion of contaminated food products or injection drug use.[3, 5]

The surgical centre undertaking the procedure must have an established TSE protocol in place for isolation and cross-contamination prevention measures. Once the medical procedure has been completed, instrument sets and associated consumables must be isolated using contaminated waste procedures and then quarantined for subsequent destruction using registered disposal agencies. Good communication and planning between clinical teams and the SSD helps ensure that correct procedures are followed. Using a track and trace system helps maintain instrument accountability, supports safe handling after use, and prevents migration of instruments between sets.

National guidance must be consulted to develop operational policies and protocols for prion risk management.

Given the nature of the causative agent of TSEs, one approach to reducing the risk of cross-contamination is to remove residual bio-contamination. Manual cleaning processes cannot be validated or reproducibly applied every time they are performed. Therefore, the use of a cleaning procedure that uses an automated instrument washer disinfector (AWD) with a validated process is a preferred option. Such processes typically use alkaline detergents with high efficiency for protein removal. The development, validation, and routine control of cleaning processes is described in ISO 15883.[7]

Despite the expectation that AWDs provide highly effective cleaning, published studies have shown that residual protein can remain on the surfaces of medical devices following AWD processing.[8, 9, 10] This suggests

that residual protein monitoring protocols are an important component of cleaning process assurance. Recommendations for acceptable levels of residual protein following cleaning can be found in standards and local guidance documents.[5, 7]

Proteins that have dried onto instrument surfaces are significantly more difficult to remove than fresh deposits.[3,11] Therefore, consideration should be given to methods that prevent tissue debris and fluids from drying during transport from the point of use to the decontamination department. Several commercially available products assist in this objective by maintaining instruments moist prior to cleaning.

Learning Objective 4: Recognize limitations of sterilization technologies and how monitoring systems affect prion risk management in sterile processing.

Steam sterilization processes have been shown to provide limited reduction of prion infectivity under certain conditions, and enhanced steam cycles are referenced in some international guidance as part of broader risk-management frameworks.[3, 6] Where such measures are considered, they are implemented based on national policy and expert guidance, not routine sterile processing practice. However, caution should be exercised to avoid a false sense of security. Evidence shows that steam sterilization may reduce prion infectivity, but high residual protein remaining on instrument surfaces after cleaning significantly compromises the effectiveness of any sterilization process. The adage "if it isn't clean, it cannot be sterilized" is highly pertinent in this context.

Some low-temperature sterilization processes have been tested for prion reduction capability. Processes that use alkylating agents, such as ethylene oxide, or low-temperature steam with formaldehyde, have been shown to be ineffective. In contrast, processes that employ oxidizing agents, such as hydrogen peroxide vapour, have demonstrated some reduction in prion infectivity under controlled experimental conditions.[12] Manufacturers' claims regarding prion inactivation should be critically examined, and the data supporting such claims should be carefully evaluated.

Monitoring of sterilization processes is addressed in multiple standards and guidance documents. In the context of prion risk management, routine monitoring systems employed for standard exposure periods may not be applicable when alternative or enhanced cycles are referenced in guidance. Chemical and biological indicators do not detect or confirm prion inactivation; however, they play an important role in verifying that validated process parameters, such as time, temperature, and sterilant exposure, have been achieved as intended. [3, 4, 5] For CRCST professionals, appropriate selection, use, and interpretation of indicators support process verification, documentation, traceability, and quality assurance by confirming consistent system performance and identifying deviations from established parameters in accordance with facility policy and national guidance.

Conclusions

CJD and other TSEs are invariably fatal and, despite being rare, pose a unique challenge to SSDs due to the resistance of prions to conventional decontamination methods. After the vCJD outbreak in the U.K., many countries reviewed their national guidance and issued recommendations to support healthcare facilities and SSDs in managing TSE-related risks.

Destruction of contaminated items by incineration provides maximum risk reduction where prion contamination is suspected or confirmed. Validated cleaning processes offer a practical approach to reducing, but not eliminating, risk. The prevention of residual tissue contamination drying before being decontaminated ensures the cleaning process is more effective.

It is vital that the SSD has a policy for dealing with potential TSE contamination and regularly reviews it in light of updated national guidance and emerging published evidence. Although it cannot confirm reduction of prion infectivity, routine monitoring, using mechanical data, chemical indicators, and biological indicators, supports process verification, documentation, traceability, and compliance. **HPN**

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Creutzfeldt-Jacob Disease and Other Prion Diseases - Practice Quiz

1. **CJD is the abbreviation for which prion disease?**
   A. Creutzfeldt-Jacob Disease
   B. Craner-Johannes Disease
   C. Coldman-Jugerand Disease

2. **CJD is predominantly a disease of which organ?**
   A. The kidneys
   B. The lungs
   C. The brain

3. **CJD is one of a group of diseases called:**
   A. Transportable Sporadic Encephalitis
   B. Transmissible Spongiform Encephalopathies
   C. Transient Soporific Endocrinology

4. **CJD is caused by**
   A. Bacteria
   B. Viruses
   C. Misfolded proteins

5. **CJD can be treated by:**
   A. Antibiotics
   B. Antiviral drugs
   C. There is no treatment

6. **What makes prions challenging for sterile service departments?**
   A. They multiply rapidly on instrument surfaces
   B. They are resistant to routine cleaning and sterilization methods
   C. They can be identified by visual inspection

7. **Instruments should be:**
   A. Allowed to dry before transportation to the decontamination department
   B. Washed under a tap before being transported to the decontamination department
   C. Kept moist and transported as quickly as possible after use to the decontamination department

8. **The presence of residual prion protein on instrument surfaces can be detected by**
   A. Looking at the instrument.
   B. Checking the weight of an instrument after cleaning
   C. Using a highly sensitive protein detection system

9. **According to international guidance, which statement best reflects current understanding of prion decontamination?**
   A. Standard sterilization cycles reliably eliminate prions
   B. No single method guarantees complete removal of prion infectivity
   C. Chemical disinfection alone is sufficient to manage prion risk

10. **Why is timely communication between clinical areas and the sterile service department important when prion risk is suspected?**
    A. To allow SSDs to make a diagnosis
    B. To ensure potential exposure risks are identified and instruments are managed according to facility policy
    C. To replace indicators and documentation requirements

11. **Which practice presents the lowest risk of patient-to-patient transmission when prion exposure is known or suspected?**
    A. Use of single-use instruments with quarantine and disposal of reusable sets
    B. Reprocessing reusable instrument sets using standard cycles
    C. Extended cleaning of reusable instruments followed by routine sterilization

12. **According to the section, how are standards and guidance on prion risk management typically used by healthcare systems?**
    A. Implemented identically worldwide without modification
    B. Replaced entirely by facility-specific procedures
    C. Used as a foundation and adapted based on national guidance and assessed risk

13. **Why can't conventional cleaning, disinfection, and sterilization processes be relied upon to fully manage prion contamination?**
    A. Because prions are primarily transmitted through airborne exposure
    B. Because prions multiply during routine sterilization cycles
    C. Because prions are resistant to many conventional reprocessing methods

14. **Which situation represents a documented example of iatrogenic transmission of a prion disease relevant to sterile processing?**
    A. Reuse of inadequately decontaminated intracranial EEG electrodes previously used on a CJD patient
    B. Consumption of beef from cattle affected by bovine spongiform encephalopathy
    C. Inherited mutation of the prion protein gene.

15. **Which tissue type is considered highest risk for prion infectivity?**
    A. Muscle tissue
    B. Gastrointestinal tissue
    C. Brain and spinal cord tissue

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