CREUTZFELDT-JAKOB DISEASE (CJD)
LEARNING OBJECTIVES

1. Define the term “prion” and identify common transmissible spongiform encephalopathy diseases, symptoms, and diagnosis challenges

2. Review the significant challenges involved with processing instruments that are or may be contaminated with transmissible spongiform encephalopathy materials

3. Discuss non-recommended and potentially useful alternatives for processing instruments known or suspected to have been used in Creutzfeldt-Jakob Disease and transmissible spongiform encephalopathy surgical procedures

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CREUTZFELDT-JAKOB DISEASE (CJD), A TRANSMISSIBLE SPONGIFORM encephalopathy (TSE), is named after the German psychiatrists who first described the disease in the 1920s. Note: “Encephalopathy” is a general term relating to brain disease, damage or malfunction. TSEs are more popularly referred to as prion diseases: a group of rare degenerative brain disorders that can be seen as tiny holes that give the tissue a “spongy” appearance when it is viewed under a microscope.

This lesson provides a foundation of background information helpful to Certified Instrument Specialist (CIS) technicians as they use special procedures to process instruments used to treat patients known or suspected to be infected with a TSE. It provides an overview of CJD, applicable healthcare policy considerations, recommended cleaning and sterilization processes, and new technologies to diagnose the disease.

OBJECTIVE 1: DEFINE THE TERM “PRION” AND IDENTIFY COMMON TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHY DISEASES, SYMPTOMS, AND DIAGNOSIS CHALLENGES

A prion is a protein-containing element that is the smallest known infectious disease-causing agent. It is not a bacteria, fungus or virus, and it contains no genetic material. For years, many researchers believed TSEs were caused by “slow viruses.” In fact, no viruses could ever be isolated, and abnormally-folded prions are now thought to be responsible for the family of TSEs in which CJD is the most widely known. The Centers for Disease Control and Prevention (CDC) indicates there were approximately 400 CJD deaths in the United States in 2010.¹

Other TSEs include Fatal Familial Insomnia (FFI), Gerstmann-Strassler-Scheinker Syndrome (GSS), New Variant Creutzfeldt-Jakob Disease (vCJD), and kuru. Prions damage specific areas of the brain (depending on which TSE causes the infection) and result in the decline of the victim’s mental abilities.

TSEs are progressive and always fatal diseases. Their incubation time can be many years (decades), and fatality is almost certain within several months after initial onset of the symptoms. Diagnosis is complicated because of spongiform changes caused by the
Specific TSE that can affect different areas of the brain at the same time.

Other challenges arise because of astrogliosos, which can be caused by several factors other than a TSE. Note: the term “astrogliosis” refers to an abnormal increase in astrocytes (cells that help keep neural tissue healthy by surrounding and providing support for and insulation between neurons in the brain). When this condition affects neurons close to the infection site, there is a reduction of inflammatory reaction that makes diagnosis difficult. These degenerations of brain functions through the loss of healthy spongiform and neurons can be seen as the patient’s mental health decreases. Examples of symptoms include possible dementia, tremors and spasticity (a muscle control disorder caused by an imbalance of signals from the central nervous system).

TSEs affect animals, as well as humans, and there is some evidence of a link between the two. Consider Mad Cow Disease [formal name: bovine spongiform encephalopathy (BSE)]. One may have been responsible for the other in a 1996 European outbreak of vCJD. In that instance, several people who developed vCJD were believed to have been infected by consuming beef products contaminated with BSE. About that same time, there were also three cases of CJD in the United Kingdom that may have been caused by an infected blood donor.

Variant CJD has a median age of death of 28 years and an illness duration period of 13-14 months (compared to CJD with a median death age of 68 years and an illness of 4-5 months) after observable symptoms. Variant CJD appears to have more symptoms and is more easily detectable then CJD. It is initially evidenced by psychiatric and sensory symptoms, followed by other neurological symptoms and progressive cognitive impairment. Magnetic resonance imaging (MRI) brain scans may enable non-invasive opportunities to diagnose the disease; however, the incubation period for vCJD is still unclear (it might be years or decades).

**OBJECTIVE 2: REVIEW THE SIGNIFICANT CHALLENGES INVOLVED WITH PROCESSING INSTRUMENTS THAT ARE OR MAY BE CONTAMINATED WITH TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHY MATERIALS**

It is important to know how to effectively process TSE-contaminated items, and strategies to do so are very difficult because they present particular barriers to most standard processing protocols.

Healthcare facilities must plan and implement an applicable policy and applicable training to manage CJD or other TSE cases that arise. These are critical to best ensure that Operating Room (OR) and Central Service (CS) staff will know what to do before the patient is in the OR.

One of the biggest problems with prion-based diseases is that once instruments are contaminated with the TSE, it becomes very difficult to clean them in a way that ensures the process has been effective. There is evidence that neurosurgical instruments contaminated with affected central nervous system tissue from implants or products (examples: dura mater and corneal grafts, and pituitary tissue) can transmit CJD if they are not properly cleaned. Although transmission of CJD in healthcare settings is exceedingly rare, CS personnel should be aware of the potential for transmission from patient to patient by contaminated instruments or equipment or by contaminated tissues.

These uncertainties are a primary
reason why many people favor using disposable instrumentation in actual or potential cases of CJD and other TSEs, and this should be considered as facilities develop their policies.

Existing loaner policies must also be re-examined. Instruments and sets must be properly cleaned and sterilized when they are received, but what procedures should be used before they are returned? Facility policies frequently require that loaners leaving facilities be decontaminated but not sterilized before being returned to their owners; however, even when this occurs, it might be preferable to decontaminate them once again before they are sterilized in the facility where they are used.

The review of and changes to existing loaner policies provide opportunities to provide additional training and education to CS, OR and even vendor personnel about CJD- and TSE-related concerns. Stakeholders should understand the policies used by the facilities previously using the instruments and how and why the loaners were processed in specific ways before their return to the lender. Note: loaners entering the facility are usually owned by the manufacturer who, in turn, may desire input to their handling and processing procedures. This is reasonable because they currently provide inservice processing and handling information to many CS personnel in many facilities.

The use of disposable instruments is a tactic to reduce problems with the management of CJD-infected instruments, and information learned in the early 2000s may be useful. The United Kingdom, along with the World Health Organization (WHO), developed guidelines for infection control management of CJD patients and the instruments and devices that come into contact with them and their tissues. In response, the Royal Melbourne Hospital in Australia began requiring the use of disposable instruments for brain biopsies.

Problems with this policy, however, involved the inability to purchase all required instruments in a disposable option, and the high cost and relative lower quality of some available disposable instruments.

Due to surgical complications in 2001 when two deaths resulted from surgical complications, the facility began using high-temperature autoclaving for 18 minutes as the standard procedure for neurosurgical instruments although the procedure is harsh on instruments.2

OBJECTIVE 3: DISCUSS NON-RECOMMENDED AND POTENTIALLY USEFUL ALTERNATIVES FOR PROCESSING INSTRUMENTS KNOWN OR SUSPECTED TO HAVE BEEN USED IN CREUTZFELDT-JAKOB DISEASE AND TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHY SURGICAL PROCEDURES

There are ongoing instrument cleaning problems and difficulties involving CJD and TSE for which solutions are needed. Fortunately, fewer than 1% of CJD cases have resulted from healthcare-associated transmission, and the majority of these transmissions have resulted from the use of contaminated tissue or grafts.3

The Joint Commission (TJC) issued an alert in 2001 that referred hospitals to the WHO guidelines and directed them to establish policies to deal with the possibility of CJD transmission. The alert and guidelines relate to possible cases of patients with unconfirmed CJD and possible tactics that might be used to avoid holding instruments in quarantine until a definitive diagnosis is determined. Doing so is typically very expensive (a neurosurgical instrument set can cost tens of thousands of dollars), removes instruments from rotation, and can delay other cases if the quarantine involves specialty instruments. There also may be concerns that quarantined instruments may be reused if they are returned to the facility’s active instrument supply.2

There are numerous alternatives for managing instruments potentially or actually exposed to CJD, ranging from autoclaving to soda lye to incinerating the devices; however, CS professionals are committed to zero tolerance for ineffective processing, and this philosophy provides the motivation to continuously seek the best processes for managing instruments known or suspected to have been used on CJD patients.

A primary instrument processing challenge for the prions associated with TSEs, including CJD, is that they are unusually resistant to conventional physical and chemical decontamination methods. Also, the prions attempt to bind to the surfaces of the instruments, making it even more difficult to remove them.
physical and chemical decontamination methods. Also, the prions attempt to bind to the surfaces of the instruments, making it even more difficult to remove them. While some enzymes have been shown to be effective, others increase the prions’ resistance to subsequent inactivation by steam sterilization. These alkaline and enzymatic detergents can reduce protein contamination, but are ineffective in eliminating the prions.

A chemical cleaner containing chlorine and sodium hydroxine (lye) provides consistent prion inactivation; however, it is corrosive and, therefore, unsuitable for many devices. Researchers continue their efforts to develop a detergent that can effectively inactivate the prions without instrument damage, and its discovery would represent a significant breakthrough in processing protocols. One current recommendation is to ensure instruments are kept damp during transport from the OR to time of decontamination, which should occur as soon as possible.

Over time, new and updated contradictions and recommendations emerge. For example, researchers now know that some sterilization and high-level disinfection processes are ineffective in deactivating prions. These ineffective processes include:

- Standard gravity and pre-vacuum sterilization cycles of 15 minutes at, respectively, 25°F (121° C) and 4 minutes at 270°F (132° C)
- Dry heat sterilization cycles, ethylene oxide, earlier forms of Sterrad (Sterrad Kos) sterilization, and radiation, microwave, and ultraviolet (UV) treatments.
- High-level disinfection (HLD) because many HLD solutions are aldehyde-based and can bind the prions more securely to the instruments.

In contrast, some processes are effective while causing less instrument damage:

- Extended gravity steam sterilization cycles for 1 hour at 250°F (121° C) and pre-vacuum cycles for 18 minutes at 275°F (135° C).
- Newer processes using radio frequency gas plasma, and vaporized hydrogen peroxide (1.5-2. mg per L).

It would also be helpful to confirm whether a patient has CJD or another TSE disease before surgery is performed. Then OR and CS personnel could determine the necessary type of instruments (reusable or disposable, when appropriate) and processing protocols without waiting for a biopsy result.

Interestingly, a new technology is emerging that may make this possible. Researchers are developing a nasal test to diagnose CJD, which would enable results to be known much more quickly and less invasively than a brain tissue biopsy.¹

**IN CONCLUSION**

There is hope that researchers will discover more effective ways to clean and sterilize instruments and/or to make available their cost-effective and appropriate quality disposable counterparts. These advancements will yield significant improvements in the care of patients with CJD and other TSE diseases.

**REFERENCES**

2. www.patient.co.uk
3. Infection Control & Hospital Epidemiology Feb 2010 Vol. 31 No 2.
1. Transmissible spongiform encephalopathy (TSE) diseases are more popularly referred to as ______ diseases.
   a. Neurological
   b. Prion
   c. Mad Cow
   d. Brain malfunction

2. A prion is which of the following?
   a. Bacteria
   b. Fungus
   c. Virus
   d. None of the above

3. Transmissible spongiform encephalopathy (TSE) diseases are always fatal.
   a. True
   b. False

4. Which is true of astrogliosos?
   a. It kills red blood cells
   b. It spreads prions
   c. It reduces inflammatory reactions close to the infection site
   d. It quickens the degeneration of neural tissue
   e. None of the above

5. Which is true of variant CJD (vCJD) compared to CJD?
   a. It has a lower median death age
   b. Illness duration is longer after observable symptoms
   c. It is more difficult to detect
   d. A and B above
   e. None of the above

6. Which is not an example of central nervous system tissue?
   a. Ganglion tissue
   b. Dura matter graft
   c. Corneal graft
   d. Pituitary tissue

7. Transmission of CJD in healthcare settings is extremely rare.
   a. True
   b. False

8. Which is a possible disadvantage in quarantining instruments potentially infected with transmissible spongiform encephalopathy (TSE)?
   a. Affected instruments are typically very expensive
   b. Instruments are taken out of rotation
   c. Affected instruments may accidently be reused
   d. All the above

9. Which is true about the prions associated with transmissible spongiform encephalopathy (TSE) including CJD?
   a. They are unusually resistant to conventional physical decontamination
   b. They are unusually resistant to conventional chemical decontamination
   c. They will bind to instrument surfaces and become more difficult to remove
   d. All the above

10. Which statement is correct?
    a. All enzymes destroy prions
    b. Alkaline detergents reduce protein contamination and eliminate prions
    c. Enzymatic detergents are ineffective in eliminating prions
    d. None of the above are correct

11. Which is true about chemical cleaners containing chlorine and sodium hypochlorite (lye)?
    a. They should be used to process instruments suspected of prion contamination
    b. These cleaners are corrosive to and not suitable for many instruments
    c. These cleaners are useful for instruments removed from quarantine
    d. These cleaners are useful if instruments remain damp after Operating Room use

12. Which process is ineffective in treating prions?
    a. Dry heat sterilization cycles
    b. Ethylene oxides
    c. High-level disinfection
    d. All the above are ineffective

13. Extended gravity steam sterilization cycles may be effective in deactivating prions while causing less instrument damage when operated for _______, at a temperature of _______.
    a. 1 hour; 250°F (121°C)
    b. 1 hour; 275°F (135°C)
    c. 18 minutes; 270°F (132°C)
    d. 15 minutes; 250°F (121°C)

14. Pre-vacuum steam sterilization cycles may be effective in deactivating prions while causing less instrument damage when operated for _______, at a temperature of _______.
    a. 1 hour; 250°F (121°C)
    b. 1 hour; 275°F (135°C)
    c. 18 minutes; 275°F (135°C)
    d. 15 minutes; 250°F (121°C)

15. Which process may be effective in deactivating prions, while causing less instrument damage?
    a. Radio frequency gas plasma
    b. Vaporized hydrogen peroxide
    c. Both A and B above

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