

## CJD – the Hidden Danger

Copyright 2015, Sterile Processing University, LLC, All Rights Reserved.

This material may not be copied or used without permission of the author.

Nancy Chobin, RN, AAS, ACSP, CSPDM

\*\*This in-service has been Approved by the CBSPD, Inc. for 1 CEU.

### Objectives:

- To discuss prions and their role in Creutzfeldt-Jakob disease (CJD)
- To review the methods of transmission of prions
- To discuss the Rutula -Weber Recommendations “Creutzfeldt-Jakob Disease: Recommendations for Disinfection and Sterilization.” - May, 2010.

**What is CJD?** Creutzfeldt-Jakob disease (CJD) is a rare, fatal brain disorder which causes rapid, progressive neurological deterioration. It has an incidence in the United States of approximately 1 case per million per year. CJD is caused by a proteinaceous infectious agent, or prion. The term “prion” arose from the words “protein” and “infectious. It causes microscopic vacuoles in the neurons of the brain. The brain becomes and appears “sponge like”.

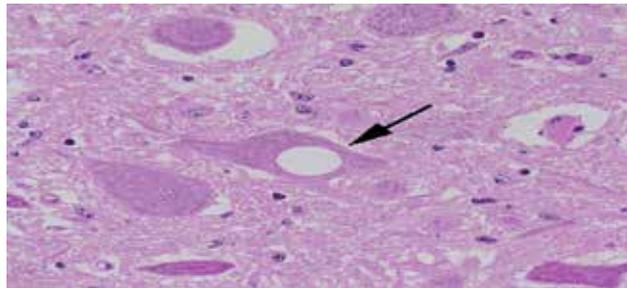


Photo Above - Brain tissue with vacuoles

“CJD affects about one person in every one million people per year worldwide; in the United States there are about 300 cases per year. CJD usually appears in later life and runs a rapid course. Typically, onset of symptoms occurs about age 60, and about 90 percent of individuals die within one year. In the early stages of disease, people may have failing memory, behavioral changes, lack of coordination and

visual disturbances. As the illness progresses, mental deterioration becomes pronounced and involuntary movements, blindness, weakness of extremities, and coma may occur.”<sup>1</sup>

According to the National Institute for Neurological Disorders and Stroke, there are three major categories of CJD:

- “In sporadic CJD, the disease appears even though the person has no known risk factors for the disease. This is by far the most common type of CJD and accounts for at least 85 percent of cases.
- In hereditary CJD, the person has a family history of the disease and/or tests positive for a genetic mutation associated with CJD. About 5 to 10 percent of cases of CJD in the United States are hereditary. Thought to be responsible for increased numbers in some communities in Chile and Czechoslovakia.
- In acquired CJD, the disease is transmitted by exposure to brain or nervous system tissue, usually through certain medical procedures. There is no evidence that CJD is contagious through casual contact with a CJD patient. Since CJD was first described in 1920, fewer than 1 percent of cases have been acquired CJD.”<sup>2</sup>

CJD usually has a long incubation period before symptoms appear. In some cases, the incubation period may be as long as 50 years. It was first identified and described in 1920 by two German psychiatrists for whom the disease is named. There are several forms of CJD.

- TSE - transmissible spongiform encephalopathy - term used to describe all fatal degenerative diseases characterized by sponge-like effect on the brain
- CJD- Creutzfeldt-Jakob Disease
- vCJD - variant strain of CJD
- BSE - Bovine spongiform encephalopathy “mad cow disease”
- Scrapie (sheep and goats)
- Transmissible mink encephalopathy
- Chronic wasting disease (elk, mule, deer)
- Feline spongiform encephalopathy

---

<sup>1</sup> National Institute of Neurological Diseases and Stroke: CJD Fact Sheet.

<sup>2</sup> National Institute of Neurological Diseases and Stroke: CJD Fact Sheet.

Prion proteins occur in both a normal form, which is a harmless protein found in the body's cells, and in an infectious form, which causes disease. The prion is virus-like but do not fit into any known category of microbe.

**What is CJD and vCJD?** The variant form of CJD (vCJD) was identified in 1996 in United Kingdom. This form of CJD was different because it was transmitted through the food chain (infected cows – hence the term “Mad Cow Disease”. It is distinguishable from the sporadic type because it occurs earlier in life. There have been no cases in US to date. Original predictions were that 100,000 cases would result from the outbreak in Europe. It is now believed that most likely 900-12,000 cases may develop.

**How is CJD Transmitted?** CJD cannot be transmitted through the air or through touching or most other forms of casual contact. Exposure to brain tissue and spinal cord fluid from infected individuals should be avoided to prevent transmission of the disease through these materials.

In some cases, CJD has spread to other people from grafts of dura mater (a tissue that covers the brain), transplanted corneas, implantation of inadequately sterilized electrodes in the brain, and injections of contaminated pituitary growth hormone derived from human pituitary glands taken from cadavers. Transmission via these routes (that are linked to medical procedures) are called *iatrogenic cases*. Since 1985, all human growth hormone used in the United States has been synthesized by recombinant DNA procedures, which eliminates the risk of transmitting CJD by this route. But what about patients we treat who had these procedures before 1985?

Many people are concerned that it may be possible to transmit CJD through blood and related blood products such as plasma. Scientists do know that, even though millions of people receive blood transfusions each year, there are no reported cases of someone contracting CJD from a transfusion. Even among people with hemophilia, who sometimes receive blood plasma concentrated from thousands of donors, there are no reported cases of CJD.

**Symptoms of CJD:** “CJD is characterized by rapidly progressive dementia. Initially, individuals experience problems with muscular coordination; personality changes, including impaired memory, judgment, and thinking; and impaired vision. People with the disease also may experience insomnia, depression, or unusual sensations. CJD does not cause a fever or other flu-like symptoms. As the illness progresses, mental impairment becomes severe. Individuals often develop involuntary muscle jerks called myoclonus, and they may go blind. They eventually lose the ability to move and speak and enter a coma. Pneumonia and other infections often occur in these individuals and can lead to death.”<sup>3</sup> One of the difficulties in diagnosing CJD is that it mimics Alzheimer’s disease or stroke.

**Diagnosis of CJD** - The only way to confirm a diagnosis of CJD is by brain biopsy or autopsy.

**Treatment** - Currently there is no treatment, the disease is 100% fatal. There is little research being done due to the low incidence of the disease.

---

<sup>3</sup> National Institute of Neurological Diseases and Stroke: CJD Fact Sheet.

**General Precautions** - There should be precautions for all patients with known or suspected prion disease and for those at high risk for the development of a prion disease. Therefore, there should be precautions taken for patients with:

- Rapidly progressive dementia
- Possible Creutzfeldt-Jakob disease (CJD)
- Gertsman-Straussler-Scheinkler (GSS)
- Fatal familial insomnia (FFI)
- Variant Creutzfeldt-Jakob disease (vCJD)
- Recipients of human growth hormone, gonadotrophin or human dura mater grafts (all from brain).
- In addition, any patient admitted for a brain biopsy without a lesion present should be suspect for CJD.

Another consideration is loaner instrumentation received for spinal or neurological cases. We have no information where the instruments were used or how they were processed. Therefore as an extra precaution, all neuro/spine loaner instruments should be considered suspect.

**Background** - The Association for the Advancement of Medical Instrumentation has adopted the recommendations of Dr. William Rutula and Dr. David Weber. They are provided in AAMI ST-79, Annex "C" as a reference.

In the late 1990's, the Joint Commission issued a Sentinel Event Alert after a patient was diagnosed with CJD. The instruments used on the index patient had not been isolated or given any special processing.

**FACTS** – The Joint Commission issued a Sentinel Event Alert for CJD in 2001 and 2013

#### **Sentinel Event Alert: 9-18-2013**

"The Joint Commission would like to clarify the recommendations in *Sentinel Event Alert #20: Exposure to Creutzfeldt-Jakob Disease (CJD) regarding the recommended practice of quarantining equipment:*

To minimize the possibility of using neurosurgical instruments that have been potentially contaminated during procedures performed on patients in whom CJD is later diagnosed, health care facilities should consider using the specific evidence-based sterilization guidelines outlined by the Centers for Disease Control and Prevention (CDC), the World Health Organization (WHO), **or the American National Standards Institute (ANSI)/Association for the Advancement of Medical Instrumentation (AAMI) ST-79:2010 Annex C.**"

This alert was based on two incidents where the neurosurgical instruments used on suspect patients were not properly sterilized after use.

**Facts on Transmission** - Transmission of CJD depends on the dose and route of entry of the prions. There have been no cases of growth hormone transmission or corneal transmission after newer methods of purifying hormones and screening of tissue occurred in US. Prior to the screening, CJD could have been transmitted through these high risk tissues.

**Tissue and Expected Concentration of CJD Agent.**<sup>4</sup> - At this time, the following types of tissue are considered high risk for prion contamination:

**HIGH RISK** – brain (including pituitary), spinal procedures, dura mater, posterior eye tissue (including retina and optic nerve)

**MEDIUM RISK** - CSF, kidney, liver, lymph node, spleen, (WHO - lung, placenta)

**LOW TO NO RISK** - Blood, urine, adrenal gland, feces, heart, bone marrow, muscle, nasal mucus, peripheral nerves, saliva, gingiva, sputum, tears

Currently, there are no known cases of transmission from a patient to a health care worker, including neurosurgeons, nurses or morticians

**What Precautions Are Needed?** - Patients scheduled for spinal cord, posterior eye (e.g. Vitrectomy) or brain procedures, including pituitary, (high risk tissues). But how do we identify if these patients have a history of CJD? Some facilities use the American Red Cross blood screening and require a pre-operative assessment, which becomes a permanent part of the medical record.

**Action to be Taken** – All facilities should have a Policy and Procedure for Handling Instrumentation and Devices from Known or Suspect CJD Patients. If you do not have one, one should be developed in conjunction with Infection Prevention using the AAMI document as the basic reference. AAMI standards are considered national standards. There should be educational programs for the surgeons and staff to understand the need for the policy.

Once the policy is implemented, there should be a system to monitor for compliance with the policy. The Policy should be assessed and refined as needed as scientific evidence changes.

Establish a Committee with representation from

- Infection Prevention

---

<sup>4</sup> AAMI. Comprehensive Guide to Steam Sterilization and Sterility Assurance in Health Care Settings, ST-79, 2013, Annex "C".

- Risk Management
- Materials Management
- Perioperative Services
- Sterile Processing/Central Service

Some of the Policy considerations include;

- How will patients be identified in the OR?
  - History?
  - Patient screening tool?

Remember, you only need to focus on patients with a history of CJD or who are having high risk surgery (brain, spinal cord, posterior eye). Also understand that most patients/surgeons are not aware of CJD history. Therefore we are very vulnerable to missing a suspect patient.

Once a known or suspect patient has been identified, there should be a method for communication of those patients. Your policy should include screening for any patient admitted for brain biopsy without lesion is suspect (if no lesion why the biopsy???).

If a Pre-op screening form is used, it should include specific questions regarding family history, blood transfusions, travel to UK and Europe during specific time periods. Screening patients in the Pre-Admission area can be most successful.

**Identifying instruments** used on known or suspect patients is essential. OR personnel should place all devices/instruments requiring special prion processing in rigid containers per OSHA regulations for transfer of contaminated instruments/sharps. The container should be labeled biohazard. There should be a means of identifying the instruments used on the high risk tissue (e.g. a special label affixed to the container, bin or red-bag (for non-sharps)).

In the OR, instruments should be treated with enzyme foam. Cover instruments with a towel moistened with sterile water (not saline) to keep soils moist. Instruments should immediately be sent to SPD for reprocessing.

In SPD, instruments should immediately be decontaminated (delays in cleaning can impact on reduction of prions). Carefully remove instruments/devices from their container/bin/bag. Make sure all instruments are opened, disassembled for cleaning. Use a method of identifying instruments requiring "Special Prion Processing" inside the basket (e.g. a colored autoclaveable tag). Unless otherwise directed by the surgical instrument/device manufacturer, no special cleaning protocols are required.

**The major change for SPD** is sterilization of the instruments. The current recommendations (AAMI ST-79) are to steam sterilize as follows:

- Pre-vacuum steam 270°F, 28-30 psig (preferred)
  - 18 minutes sterilization time
- Gravity displacement steam 250°F, 15-17 psig
  - 60 minutes sterilization time

It is important to understand that all standard methods of sterilization or high level disinfection appear to be ineffective for CJD. These include alcohol, glutaraldehyde, boiling, hydrogen peroxide, detergents iodophors, dry Heat, ionizing/UV radiation, EO, peracetic acid, formaldehyde, phenolics, formalin, and routine steam sterilization cycles.

The World Health Organization has published different recommendations for prion inactivation. These include sterilize all devices for all cases for 18 min at 270°F, expose surfaces to 1N of NAOH(caustic/corrosive) for 30-60 min and exposure of surfaces to NaCL(bleach) for 30-60 minutes. Unfortunately exposure of surgical instruments to sodium hydroxide or bleach will destroy the surgical instruments.

There is concern about the ability to effectively clean lumened devices. Therefore for known or suspect CJD cases, it is recommended that the OR use disposable suctions, have available disposable craniotomy sets and covers for power equipment.

**Special Considerations** – Since immediate use steam sterilization (IUSS or flash) is ineffective for prions, IUSS should not be used for any known or suspect instrumentation. Instruments or devices that can only be processed in low temperature sterilization or ethylene oxide should be discarded. These methodologies are INEFFECTIVE against prions.

**Loaner Instrumentation** – If your facility uses loaner instrumentation or devices for high risk tissue (e.g. spine, posterior eye), the loaner instruments should be considered suspect. Therefore these loaner instruments should, upon receipt from the representative be;

- Decontaminated per the instructions for use– pay particular attention to lumened devices
- Sterilized, unwrapped using the prion inactivation cycles recommended.
- Wrap and re-sterilized per manufacturer’s instructions.

**NOTE:** Unless the instrument/device manufacturer has provided written instructions for CJD or prion inactivation cycles, you should contact the manufacturer for guidance. SPD personnel are not permitted to exceed the manufacturer’s IFU for sterilization.

**Traceability to the Patient** - Contaminated items that have been in contact with high-risk tissue and have not been processed according to these recommendations (e.g., medical devices used for brain biopsy before diagnosis) should be recalled and appropriately reprocessed. A tracking system should be in place that permits recall of devices used on high-risk tissue and high-risk patients. This system should permit identification of the patient on which the devices were used, the date they were used, the procedure performed, and the surgeon's name. This can be performed by using a 2-part tag or card that is affixed to all trays that will be used on high risk tissue.

**Problem** – The CDC, FDA and World Health Organization do NOT agree on methods of inactivation of prions. This causes confusion for facilities. AAMI standards are based on what occurs in the US with prion disease therefore these standards should be used.

### **CJD - Conclusion**

Remember, the only thing which remains constant with microorganisms is their ability to change to adapt to adverse changes in their environment. If we are prepared for CDJ it may not get an upper hand. When the Joint Commission issues a Sentinel Event Alert, this requires we take action.

Develop policies and procedures based upon good scientific evidence. Educate all personnel, including physicians. Monitor for compliance with the policy.

### **References**

AAMI. Comprehensive Guide to Steam Sterilization and Sterility Assurance in Health Care Settings, ST-79, 2013, Annex "C".

National Institute for Neurological Disorders and Stroke. Creutzfeldt-Jakob Disease Fact Sheet. [http://www.ninds.nih.gov/disorders/cjd/detail\\_cjd.htm](http://www.ninds.nih.gov/disorders/cjd/detail_cjd.htm). Accessed 1/8/15

Rutula, William and Weber, David. "Creutzfeldt-Jakob Disease: Recommendations for Disinfection and Sterilization. *Clinical Infectious Diseases: Healthcare Epidemiology*. May, 2010.

Recommendations for Processing of Devices Used on CJD Patients - American Neurological Association and the Association for Practitioners in Infection Control 1996.

Joint Commission for the Accreditation of Healthcare Organizations. Sentinel Event Alert, September, 2013.

## **See Next Page**

## Quiz on CJD

Please click on the link below to take the quiz.

[https://www.spdceus.com/ceus/cjd\\_quiz.htm](https://www.spdceus.com/ceus/cjd_quiz.htm)

Good Luck!