Purpose
The purpose of this self-learning packet is to present nurses with current information that will assist in improving care of the patient with CJD in Perioperative Services.

Objectives
Upon completion of this self-learning packet, the reader will be able to:
Define CJD.

List the types and incidence of CJD.

Describe the signs and symptoms of CJD.

Identify procedures for caring for the patient and instruments in the OR.
Instructions

In order to receive credit, you must:

- Complete the post test in this packet and sign the attestation at the bottom of the post-test.

- Submit the post test and evaluation to your manager or Nurse Educator

Introduction

Creutzfeldt-Jakob Disease (CJD) is a rare, degenerative, rapidly progressing, fatal brain disease.

The recent discovery of Mad Cow Disease in the US highlighted the need for U.S. healthcare professionals to become familiar with this potentially epidemic producing disease.

The potential to transmit the disease during surgery suggests that we become intimately familiar with CJD.

CJD

CJD is named for two German Psychiatrists, Hans Gerhard Creutzfeldt and Alfons Jakob.

Originally considered a “slow viral infection”, we now know CJD is caused by infectious protein particles which lack RNA and DNA called prions.

These misshaped proteins convert normal proteins in the brain cell by changing the way they fold which, in turn, changes the way they join with other cell components. As these abnormal prions increase, healthy proteins deteriorate and infectivity increases. The incubation period is 4-30 years. Males and females are equally affected.

CJD is the most common member of the disease family called Transmissible Spongiform Encephalopathies. Other examples are Transmissible Mink Encephalopathy found on mink farms, Chronic wasting disease in US Mule Deer and Elk, and Feline Spongiform Encephalopathy in domestic cats.

Transmissible Spongiform Encephalopathies are fatal neurodegenerative diseases that affect both humans and animals. When TSE’s were first discovered, it was felt that the disease would remain in the species from which it was endemic. However, we now know that it can be passed from one species to another. The brain continues to be damaged, has microscopic holes (like a sponge) and often is described on autopsy as looking like Swiss cheese.

TSE prions are resistant to steam sterilization, dry heat, ethylene oxide gas, and chemical disinfection.

Great Britain used to make feed for beef containing bone meal and parts of sheep. Scrapie in sheep is a TSE. In 1986 it was noticed that some cows began to stagger, wobble and appear fearful. This was the beginning of Bovine Spongiform Encephalopathy (BSE) or Mad Cow Disease.

At the time approximately 200,000 cattle were affected; now we hear only of an occasional outbreak. When this disease is transmitted to humans, we call it CJD.

Americans became aware of the disease in 1983 when the famous choreographer, George Balanchine, died of this disease.
There are 5 types of CJD. Variant CJD appeared in the 1990’s in humans consuming the afore mentioned cattle. The occurrence of vCJD has increased 23% since its recognition in 1996. Great Britain made it illegal to eat certain parts of animals, especially nerve tissue such as brains and spinal cord. Gelatin, which is a beef byproduct, and can be in cosmetics, desserts and medication capsules, became questionable. The age group developing this type of CJD disease is much younger than normal-the mean age is 29 years. Most patients develop the disease in their 50’s and 60’s. Development of the disease is much slower than normal, and it has a longer clinical course (9 months-3 years.) It has a broader tissue distribution; prions have recently been found in the adrenal gland, thymus, spleen, blood and tonsils.

Iatrogenic (icjd) is induced by medical treatment. This has happened by injection of human growth hormone, by transplanting tissues such as Dura mater or corneas, by contaminated surgical instruments, and, in a few cases, blood transfusions.

Genetic CJD occurs in 10-15% of all CJD cases and usually occurs in the 40’s and 50’s. 50% of the children of those with the mutated gene will inherit it. (autosomal dominant)

Sporadic CJD is represents approximately 90% of the cases. Incidence of this type is about 1/million and usually affects people over 50. This type is known for rapid deterioration and a mean of about 4 months between onset and death.

The last type has only been seen in the Fore tribe in Papua, New Guinea. We call it Acquired-Kuru and it is caused by cannibalism. Now that education is decreasing this practice, the cases are appearing less frequently.

The patient with CJD initially presents neglecting his personal appearance, such as bathing, grooming and/or eating. They will progress to apathy or irritability, then become forgetful, easily fatigued and disoriented. Family will report sleep disorders. They will exhibit impaired judgment and language skills, progressing to aphasia and be unable to read or write. They will be emotionally unstable and will have dizziness and tingling in lower extremities.

In a short time they will need 24 hour care. They will need assistance standing and walking, eating, drinking and have difficulty swallowing. They will have visual disturbances, hyperkinesias (decreased motor reaction to stimuli), extrapyramidal rigidity, nystagmus, tremor and eventually ataxia. They will be incontinent.

All 5 types will feature rapid, progressive mental deterioration with myoclonus (twitching or clonic spasm of a muscle or group of muscles). 70% die within 6 months; the rest within 1 year.

At some point after the patient becomes symptomatic, their EEG will show characteristic spikes which are considered diagnostic in nature. MRI and CT can be utilized to rule out other forms of dementia and reveal atrophy associated with CJD.

Diagnosis can only be made by histologic exam of affected brain tissue. Most commonly it is diagnosed during autopsy; brain biopsy may be accomplished via Craniotomy or Stereotactic brain biopsy.

There are some new tests being utilized to attempt earlier, more accurate diagnosis. The first is checking for the Genetic Marker 14-3-3 Protein Assay which is used to test a patient with a familial history to see if they have the gene. Erythrosin B Dye is used to check for effectiveness of cleaning methods. Prions have been found in tonsils, so it has been used to check for cleanliness of anesthesia equipment in reusable equipment eg: LMA’s. A urine test using dialyzed urine checks for a protease-resistant protein prion to identify carriers. One of the newest discoveries is that a tonsil biopsy can be used for a PREMORTEM confirmation of vCJD.
Some experimental drugs are being tried to treat CJD. Quinacrine, an anti-malarial, is given to some patients in the hope it will bind to the distorted prion and slow down the degeneration. Great Britain is injecting Pentosan. No effectiveness has been shown at this time.

Infectious tissue classifications must be considered when discussing CJD contamination.
High Infectivity Tissue includes the brain, spinal cord and eye. Blood, once thought to be a No infectivity tissue, is being relooked at since there have been a couple of reported cases of believed contagion from blood transfusion. Lymph nodes were considered low infectivity along with Kidney, Liver, Lung, Spleen, and Placenta. These are also being researched as prions can be stored in tonsils, as mentioned previously.

If a brain biopsy is being performed in the OR and CJD is suspected, certain precautions must be taken. The Dept. of Pathology, Infection control and Central Supply Supervisor should be notified of the case. They should be called as specimens or instruments are being sent and informed that they need special precautions. All non-essential equipment and furniture must be removed from the room. Blood and body fluid precautions must be used. All personnel must use PPE: gowns, eyewear, double gloves and masks. Hand drills and not power tools must be used to prevent aerosolization. Disposable brain biopsy kits must be used whenever possible. Red bag all garbage if patient has or is suspected to have CJD. The surgeon must regown and glove after specimen is removed.

It was previously mentioned that these prions are resistant to all forms of steam, dry heat, or EO sterilization. The chemicals we use for sterilization (eg: CIDEX) actually make the prions more stable, therefore less responsive to normal sterilization methods. NSLIJ Policy states that if reusables must be used, as in a corneal transplant tray, the instruments must be kept moist until cleaning. Instruments should be placed in a red bag after use and then in an impervious container if possible for transport. The instruments used on the high infectivity tissues should be separated from the others for decontamination. All fluids are solidified by being treated with Isolyzer. Instruments exposed to high infectivity tissues must be cleaned then steamed

- Prevac 18 min. 272 degrees
- Gravity 60 min. 270-273 degrees

After this decontamination, the instruments are wrapped and sterilized in the usual manner. The equipment in the room is cleaned with bleach post-op.

Post-operatively, the patients are handled with Standard Precautions. Urine, feces, tears, semen are all considered no infectivity tissues.

**Conclusion**

CJD is real, it is dangerous and it is here. **You** have the responsibility to inform those around you. Only through your vigilance can we limit the transmission of CJD to uninfected patients.
References:

Stricof, R.; Thomas, N.; Schonberger, L. B. An Investigation of Potential Neurosurgical Transmission of CJD: Challenges and Lessons Learned Infection Control and Hospital Epidemiology, March 2006, Vol. 27, 302-303


Scicchitano, L. Bovine Encephalopathy and CJD: Background and Implications for Nursing Practice American Society of Ophthalmic Registered Nurses, Oct-Dec 2004, 19-21


Syosset Hospital Infection Control Standardized Policy Management of Known or Suspect CJD and other Transmissible spongiform Encephalopathic Diseases.
CJD Post Test

ATTESTATION STATEMENT

Course #3200

1) I have received the Self-Learning Packet (SLP) on CJD.
2) I have read the entire packet and have had the opportunity to ask questions related to its content.
3) Return the posttest & Attestation Statement to your nurse educator.
4) Keep the SLP for your own reference.

Employee Signature: ________________________________ Date: ____________________

1-The incubation period for Creutzfeldt –Jakob disease (CJD) is:
   _____ A. One year
   _____ B. Five years
   _____ C. One to four decades
   _____ D. Five to six decades

2-Which pathologic finding is common to CJD?
   _____ A. Cerebral atrophy
   _____ B. Retinal degeneration
   _____ C. Demyelinization of nerve fibers
   _____ D. Sponge-like holes in the brain

3-The theory that a prion causes CJD indicates which pathophysiologic event occurs when the disease is manifested?
   _____ A. Healthy proteins deteriorate
   _____ B. The blood will not clot.
   _____ C. Pituitary hormone is absent.
   _____ D. Cardiac arrhythmias.

4.- Which symptoms are most common to CJD?
   _____ A. Hair loss and fatigue
   _____ B. Elevated blood sugars and ketones.
   _____ C. Diarrhea and protein deficiencies.
   _____ D. Myoclonus and problems with speech and swallowing.
5- Which diagnostic study is most likely to support the diagnosis of CJD?
   _____A.  Angioplasty
   _____B.  Blood Counts
   _____C.  EEG and Brain Biopsy
   _____D.  Lumbar Puncture

6- Two known mechanisms for the transmission of CJD is?
   _____A.  Direct skin contact and airborne transmission
   _____B.  Genetic predisposition and blood borne pathogens.
   _____C.  Corneal Transplant and contaminated surgical instruments.

7- Infectivity is found most often and in high concentration in the:
   _____A.  CSF and urine
   _____B.  Blood and saliva
   _____C.  Brain, spinal cord, and eye

8- Which type of isolation should OR workers caring for a patient with CJD use?
   _____A.  Blood and body precautions
   _____B.  Special respiratory precautions
   _____C.  Contact isolation
   _____D.  Standard Precautions

9- How should high risk critical or semi critical instruments be handled following a high tissue infectivity procedure?
   _____A.  Place instruments in a case cart unwrapped and send down to CSS.
   _____B.  Place instruments in a bag, place bag in impervious container, send in a sealed case cart marked “Biohazard” and call CSS Supervisor to alert that instruments require special precautions.
   _____C.  Place instruments in case cart that is labeled “Biohazard” and send to CSS.

10. How should post-op CJD patients be handled by healthcare workers?
    _____A.  No special requirements beyond Standard Precautions
    _____B.  Blood and Body Fluid Precautions
    _____C.  Contact Isolation
    _____D.  Special respiratory precautions
Self-Learning Packet Evaluation

CREUTZFELDT-JAKOB DISEASE

Date: ________________

Your position?

Please take a few moments to answer the following questions by marking the appropriate boxes

1) The content provided was beneficial. ☐ ☐ ☐ ☐ ☐
2) The packet met the stated objectives. ☐ ☐ ☐ ☐ ☐
3) The packet was easy to read. ☐ ☐ ☐ ☐ ☐
4) The posttest reflected the content of the packet ☐ ☐ ☐ ☐ ☐
5) The course was: ☐ Mandatory ☐ Optional

Please answer the following questions:

How long did this packet take you to complete? ____________________

What have you learned that you will apply in your work? ____________________

What was the best part of the packet? ____________________

What would you suggest be done differently? ____________________

Additional Comments:

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Thank you for your input. Comments will be evaluated for further revisions. Please return this evaluation to Nursing Education & Staff Development, with your posttest and signed attestation.