I. **POLICY:**

1. Human prion diseases, Creutzfeldt Jakob disease, (CJD, variant CJD, fatal familial insomnia, Gertsmann- Straussler syndrome) are fatal, degenerative neurological disorders.
2. Prions are a unique classification of infectious agents with a genetic component that are thought to be transmitted through direct inoculation rather than through traditional routes (example blood borne, skin contact, droplet, airborne).
3. Prion diseases are infectious. Infectivity is based primarily on laboratory studies of different prion diseases in humans and animals.
4. Prions are resistant to chemical disinfection (example alcohol, gluteraldehyde) and routine sterilization (example steam, ethylene oxide, gas plasma, peracetic acid).
5. Compliance with this policy is required to prevent cross infection of patients, to minimize environmental contamination and reduce or eliminate health care workers exposure.

<table>
<thead>
<tr>
<th>Infectivity</th>
<th>Substance</th>
</tr>
</thead>
<tbody>
<tr>
<td>High infectivity tissue</td>
<td>Central nervous system, specifically the brain, spinal cord and eye</td>
</tr>
<tr>
<td>Low infectivity tissue and fluids</td>
<td>CSF, kidney, liver, lungs, lymph nodes/ spleen and Placenta</td>
</tr>
<tr>
<td>No detectable infectivity</td>
<td>Blood, other body fluids, and tissue such as adipose, adrenal gland, gingival tissue, heart muscle, thyroid gland, etc.</td>
</tr>
</tbody>
</table>

*Basic pathology doesn’t necessarily exclude the possibility of CJD.

6. Protein 14-3-3 test performed on CSF is the test ordered when looking for CJD.
7. Patients scheduled for brain biopsy without a lesion present must be assessed for CJD by having a CJD Risk Assessment Tool/Pre-op checklist completed by the neurosurgeon/procedurist.

8. Patients should be screened for the risk of prion disease, and any information discovered should be conveyed to the Operating Room/Procedural Area during scheduling.

9. Conveying this information during scheduling provides adequate time to plan instrument use, decontamination, and discuss alternatives to the use of complex instruments and implant sets requiring reprocessing.

10. All patients who are known or suspected of having CJD require special set-ups and processing of devices used for surgical procedures and procedures involving the central nervous system, the brain, spinal cord and eye.

11. Disposable instruments and supplies will be used whenever possible.

II. PURPOSE:

1. To prevent the transmission of CJD.

2. To insure proper preparation and handling of any instrument, drill, patient care apparatus, tissue and fluid specimens, from a patient undergoing a Brain Biopsy, Craniotomy or lumbar puncture procedure with suspected CJD.

III. PROCEDURE:

OR, SPD, Procedural Areas and Laboratory personnel must be knowledgeable in special procedures necessary for handling instruments, patient care apparatus, and tissue and fluid specimens used on known or suspected CJD patients.

A. General Precautions

1. Precautions are used for all patients with known or suspected prion disease and for those at high risk for the development of a prion disease, including all patients with rapidly progressive dementia; those with possible Creutzfeldt-Jakob Disease (CJD), Gerstmann- Straussler-Scheinker (GSS), fatal familial insomnia (FFI), or variant Creutzfeldt –Jakob Disease (vCJD); recipients of human growth hormone gonadotrophin or human dura mater grafts (all from brain); and any patient who answers yes to the Risk Assessment Tool/Pre-op Check List. In addition, any patient admitted for a brain biopsy without a lesion present should be suspect for CJD.

2. The Infection Prevention Department and other departments involved with infection prevention (i.e. surgical services, central services/sterile processing, pathology and laboratory services) should be notified by clinicians when a patient with known or suspected prion disease is scheduled to undergo any invasive procedure in which there may be exposure of personnel or instruments that contain potentially infectious tissues.

3. Clinicians should notify the Infection Prevention department of all patients with a known or suspected prion disease.

4. Standard precautions should be used for all patients with known or suspected CJD.
   a. Additional precautions (i.e. contact) are not necessary.
   b. Gloves should be worn for the handling of blood and body fluids (i.e. secretions and excretions).
c. Masks, gowns and protective eyewear should be worn if exposure to blood or other material that is potentially infectious to mucous membranes or skin is anticipated.

5. Because standard decontamination of tissue samples (i.e. formalin) or specimens may not inactivate CJD, all tissue samples should be handled using standard precautions (i.e. gloves).
   a. The tissue samples and specimens should be labeled as” biohazard” and as “suspected CJD” before being sent to the laboratory.

6. No special precautions are required for disposal of body fluids.
   a. Such fluids may be disposed of by means of a sanitary sewer.
   b. Blood and blood contaminated fluids should be managed per state regulated for required medical waste.

7. Regulated medical waste (i.e. bulk blood pathological waste, microbiological waste and sharps) should be managed per state regulations.

8. Laundry should be managed as required by the Occupational Safety and health Administration (OSHA) rule on blood borne pathogens. No additional precautions are required.

9. Surgical drapes, gowns and single-use supplies should be used whenever possible and incinerated after use.
   a. Drapes and gowns that are in contact with highly infectious tissue during these procedures. Routine hospital laundry does not inactivate prions
   b. Work surfaces should be covered with disposable, impervious material that can be removed and incinerated after the procedure.

10. No special precautions are required for the handling of food utensils.

11. When a patient expires, ensure that the morgue and funeral home are notified that the patient had CJD. No excess precautions need to be taken in regard to burial (i.e. no special cemetery is needed).

12. Patients with known or suspected prion disease should not serve as donors for organs, tissues, blood, blood components or sources of tissue (i.e. dura mater and hormones).

B. Procedural Areas: Precautions for Patient’s Undergoing Lumbar Puncture with Suspected Creutzfeldt-Jakob Disease.

1. Immediately notify Infection Prevention and the Chemistry Lab with patient name & medical record.

2. All CSF specimens must be hand delivered to the chemistry lab in the appropriate bio-hazard bag.

3. Specimens for the Pathology lab must include “R/O CJD” on the handwritten requisition slip accompanying the specimen(s).

4. Dispose of all sharps in the sharps container. At the completion of the lumbar puncture, notify environmental to remove sharps container for incineration.

5. Dispose of all associated spinal tap waste/supplies in a red bag and request prompt garbage bag removal by the environmental staff.

6. Follow standard precautions – Personal Protective Equipment (gloves, face shields and gowns) must be used during procedures with potential exposure to any blood or body fluids.

C. Operating Room: Precautions for Patient’s Undergoing Brain Biopsy, Craniotomy with Suspected Creutzfeldt-Jakob Disease.
1. The scheduling of all Brain Biopsy, Craniotomy procedures require a Creutzfeldt-Jakob Risk Assessment Tool/Pre-Operative Checklist completed by the Neurosurgeon prior to the patient’s arrival in the Preoperative Care Unit.

2. The CJD Risk Assessment Tool/Quarantine Report shall be placed in the H&P section of the patient medical record and document on the Pre-procedural Checklist.

3. A YES answer to any questions on the Risk Assessment Tool/Quarantine Report requires that;
   a. The surgeon must notify the Director of Perioperative Services/Designee or the Nursing Supervisor during off shift hours.

4. The following individuals are informed by the Director of Perioperative Services/Designee or the Nursing Supervisor during off shift hours when a suspected CJD procedure is scheduled.
   a. The OR Assistant Nurse Manager/Nursing Supervisor/designee
   b. Manager of SPD
   c. Infection Prevention Director/designee
   d. The Director of Pathology or designee
   e. The Director of Laboratory Services or designee
   f. Anesthesiologist

5. Patients for surgery without a completed CJD Risk Assessment Tool/Quarantine Report due to the emergent nature of the procedure requires implementing the CJD segregation and quarantine procedure.

6. Copies of the CJD Risk Assessment Tool/Quarantine Report shall also be kept in the Peri-operative area.

7. The Surgeon/Procedurist, Anesthesiologist and RN caring for the patient will review the CJD Risk Assessment Tool/Quarantine Report in the Preoperative Care Unit/Procedural Area, or prior to the start of the procedure.

8. Labeling and Handling of Specimens
   a. All CSF specimens must be hand delivered to the chemistry lab in the appropriate bio-hazard bag.
   b. Specimens for the Pathology lab must include “R/O CJD” on the handwritten requisition slip accompanying the specimen(s).

D. PROCEDURE FOR INSTRUMENT SEGREGATION AND QUARANTINE INTRA-OPERATIVELY
1. The OR will request a designated red transfer container for high risk instruments from SPD. High risk instruments are defined as those in contact with dura mater or eyes from suspected or known CJD patients.

2. Low risk instruments are those that have not come in contact with dura mater or eyes from suspected or known CJD patients. These instruments will be processed routinely and placed in the case cart at completion of the procedure.

3. Disposable instruments and supplies shall be used whenever possible.

4. Non powered drills/saws shall be used to minimize percutaneous injury and eliminate the generation of aerosols.

5. The surgical team shall wear double gloves.

6. No hand to hand passing of instruments shall occur.

7. A neutral zone is established to minimize accidental sharps injury.

8. During the surgical procedure, high risk instruments must be segregated from the no risk instruments.
E. PROCEDURE FOR INSTRUMENT SEGREGATION AND QUARANTINE POST PROCEDURE

1. Instruments and other materials subject to re-use should be kept moist between the time of exposure to infectious materials and subsequent decontamination and cleaning. Removal of adherent particles through mechanical cleaning will enhance decontamination process.

2. At the completion of surgery, OR personnel must separate and immerse all high risk devices/instruments requiring special handling in enzymatic detergent prepared in accordance with manufacturer’s recommendation.

3. Enzymatic detergent is obtained from SPD.

4. The designated red transfer container is then closed and labeled Prion High Risk Special Handling.

5. Attach a copy of the completed CJD Risk Assessment Tool/Quarantine Report to the outside of the red container.

6. Place labeled red container on top of the case cart.

7. All other equipment shall be placed back into the case cart. The case cart is labeled Prion No-Risk.

8. All labeling must take place in the Operating Room before equipment is transferred; the labeling should be appropriately laced around the entire container.

9. Inform SPD of the transfer via extension 8568.

10. The circulating RN must ensure that the OR section of the quarantine report is complete, signed and sent with the red transfer container to the Central Sterile Decontamination Room.

11. Discard all used disposable supplies and linen from suspected or known CJD patients in a red medical waste trash bag.

12. Specimens for the Pathology lab must include “R/O CJD” on the handwritten requisition slip accompanying the specimen(s).

13. Cleaning of the Operating Room shall be in a routine fashion.

14. Return unused, unopened supplies to SPD return carts.

F. Instruments Processing for Suspected / Known CJD Patients

1. SPD must acknowledge receipt of instruments of suspected or known CJD patients by signing quarantine report. The signed report is then forwarded to the SPD Manager/designee.

2. The labeled red transfer container will be placed in a designated area and left undisturbed under quarantine.

3. All Prion High Risk reusable instruments shall remain under quarantine indefinitely with report attached awaiting pathology results.

4. All other reusable instruments shall be processed routinely in accordance with AAMI and AORN guidelines for cleaning and will be held awaiting final assembly.

5. If the pathology report is negative, Pathology will notify the SPD Manager / Designee and all instruments shall be released for sterilization in accordance with all AAMI and AORN guidelines for cleaning and sterilization.

6. If the pathology report is positive for CJD, Pathology will notify the SPD Manager / Designee and all reusable devices/instruments that have come in contact with tissue considered to be highly infective from patients at high risk for a prion disease, will be processed in accordance with AAMI standards and AORN Guidelines.
a. Clean the instrument/device with an instrument cleaner as soon as possible after use
b. Remove all liquids after cleaning the device/instrument
c. Steamed sterilize the device/instrument for eighteen minutes in a pre-vacuum sterilizer with a cycle temperature of 134 degrees Celsius (272 degrees Fahrenheit), or
d. Steam sterilize the device/instrument for 60 minutes in a gravity displacement sterilizer with a cycle temperature of 132 degrees Celsius, or.
e. Immersion of the device/instruments in 1N sodium hydroxide (NaOH), for one hour, followed by removal of instrument and water rinse
7. The device/instrument is then cleaned, wrapped and terminally sterilized by conventional means.
8. Devices/instruments that have been contaminated with medium, low, or no-infectivity tissue can be cleaned and disinfected or sterilized using conventional protocols of heat, chemical sterilization, or high-level disinfection.

G. Drills/Power Equipment
   Drills/power equipment shall not be used.

H. Loaner Equipment
   1. Loaner equipment used in Prion High-Risk cases shall be processed in the same manner as hospital owned equipment.

I. Chain of custody documentation for suspected equipment
   1. A CJD Risk Assessment Tool/Pre Checklist shall be completed by the surgeon/procedurist.
   2. Any item checked with a (yes) shall be the indicator for segregation and quarantine of surgical equipment.
   3. The original will be sent to the Pathology Lab with the specimen.
   4. A second copy of the quarantine report shall accompany all high risk equipment that will be transferred to the Central Sterile decontamination room.
   5. A third copy of the report shall remain on the patient’s chart.
   6. A fourth copy will be sent to the Laboratory for any additional tests ordered.

J. Occupational Exposure
   1. Care must be taken to avoid self-inoculation with sharp objects.
   2. Although scientifically unproven, percutaneous exposure to the CSF or brain tissue of an infected person may be followed by rinsing the wound with 0.5% sodium hypochlorite (a 1:10 dilution of bleach) for several minutes and then washing with soap and water.
   3. Mucous membrane exposure to infectious tissues or fluids should be managed by irrigating the mucous membranes thoroughly with saline for several minutes.

K. Environmental Cleaning
   1. Before the operative and invasive procedure begins, personnel should cover work surfaces with a disposable, impervious material that can be removed and surfaces decontaminated after the procedure if contaminated with high risk tissue.
   2. Special cleaning procedures should be used if environmental contamination with
high risk tissue (example brain, spinal cord, eye tissue) from a patient who is diagnosed with or suspected of having Creutzfeldt - Jakob disease (CJD) occurs.

3. Clean the non-critical environmental surface with a detergent.
4. Decontaminate the surface with a solution of either sodium hypochlorite (1:5 to 1:10 dilution with 10,000 ppm to 20,000 ppm available chlorine) or sodium hydroxide (1N NaOH).
5. Apply the disinfectant solution for a contact time of 30 minutes to 1 hour.
6. Use an absorbent material to soak up the solution.
7. Discard the cleaning material in an appropriate waste container.
8. Rinse the treated surface thoroughly with water.
9. Change into clean scrub attire when the procedure and clean-up is completed.
10. If the environment is not contaminated with high risk tissue, routine cleaning procedures should be used.
11. Regulated medical waste generated during patient care, including waste contaminated by high risk tissue that is decontaminated, should be managed in accordance with standard waste management procedures.

IV. REFERENCES:

World Health Organization Infection Control Guidelines for Transmissible Spongiform Encephalopathies. 1999


AAMI Standards 2011

VI. APPROVAL SIGNATURES

Approved by:

_________________________________________________________  July 31, 2014
Chief Administrative Officer
Saint Peter’s University Hospital

Origination Date: 1/15/02
Supersedes Date(s): 1/25/14, 7/31/14
Reviewed Date: 6/2014
Revised Date: 6/2014
Hosp Admin-950-50
CREUTZFELDT-JAKOB DISEASE (CJD) RISK ASSESSMENT TOOL

PRE-PROCEDURAL CHECK LIST/QUARANTINE REPORT

Risk assessment for identification of possible CJD patients includes answering the following questions:

1. Does the patient have known/suspected CJD or exhibit any of the classic clinical symptoms?
   - Rapid Progressive Presenile Dementia
   - Myoclonus (Myoclonic Jerks)
   - Progressive Motor Dysfunction (Dementia with Lower Motor Neuron Findings) Cerebellar Ataxia
   - Abnormal EEG patterns with periodic complexes of repetitive triphasic spikes and slow waves at 1 Hz9 Cerebellar Ataxia
   - [ ] YES [ ] NO
   - [ ] YES [ ] NO
   - [ ] YES [ ] NO

2. Is there a familiar history of CJD?
   - [ ] YES [ ] NO

3. Is there a family history of any other inheritable spongiform encephalopathy (Gertsman-Straler-Scheinker or Fatal Insomnia)?
   - [ ] YES [ ] NO

4. Has the patient ever received any human pituitary growth hormone therapy?
   - [ ] YES [ ] NO

5. Does the patient have a history of receiving a human dural enlargement?
   - [ ] YES [ ] NO

6. Does the patient have a history of receiving a human corneal transplant?
   - [ ] YES [ ] NO

7. Does the patient have a rapidly progressive dementia not yet diagnosed?
   - [ ] YES [ ] NO

8. Has the patient received blood products from a CJD donor?
   - [ ] YES [ ] NO

9. Does the patient present a quartet of dementia, myoclonus, periodic EEG activity and rapid progression?
   - [ ] YES [ ] NO

Signature of MD: ___________________________ Date: ___________________________

CREUTZFELDT-JAKOB DISEASE (CJD) QUARANTINE REPORT

<table>
<thead>
<tr>
<th>OR/PROCEDURE ROOM #</th>
<th>NAME OF PERSON RECEIVING INSTRUMENTS IN SPD:</th>
</tr>
</thead>
<tbody>
<tr>
<td>SURGEON/PROCURIST NAME</td>
<td>DATE OF INSTRUMENT QUARANTINE IN SPD:</td>
</tr>
<tr>
<td>SCRUB TECH NAME:</td>
<td>DATE OF PATHOLOGY REPORT</td>
</tr>
<tr>
<td>RN’S NAME:</td>
<td>DATE OF ADDITIONAL LAB REPORT(S)</td>
</tr>
<tr>
<td>INSTRUMENTS TRANSPORTED TO SPD BY</td>
<td>DATE PATHOLOGY REPORT SENT TO MD:</td>
</tr>
<tr>
<td>NOTIFICATION OF INFECTION PREVENTION (DATE/TIME)</td>
<td>NOTIFICATION OF LABORATORY SERVICES (DATE/TIME)</td>
</tr>
<tr>
<td>NOTIFICATION OF PATHOLOGY (DATE/TIME)</td>
<td>DISPOSITION OF INSTRUMENTS (DATE/TIME)</td>
</tr>
</tbody>
</table>

NS-373 (6/14)