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Beaumont

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Area	Laboratory-Safety
Applicability	All Beaumont Hospitals

Handling Anatomic Pathology Specimens and Autopsies with Potential Prion Disease or Transmissible Spongiform Encephalopathies (TSE) - Creutzfeldt-Jakob Disease (CJD)

Document Type: Procedure

I. PURPOSE AND OBJECTIVE:

A. The purpose of this document is to address how Anatomic Pathology handles tissue from patients suspected of having a prion disease and how to handle autopsy requests in patients suspected to have prion disease.

II. GENERAL INFORMATION:

A. Creutzfeldt-Jakob disease (CJD) is a rapidly progressive, invariably fatal neurodegenerative disorder believed to be caused by an abnormal isoform of a cellular glycoprotein known as the prion protein. Exposure to brain tissue, spinal cord fluid and possibly blood from infected persons should be carefully handled to prevent transmission of the disease through these materials.

B. **For Tissue Specimens**: Whenever clinical suspicion of a prion disease is suspected, Corewell Health Laboratory will initially send tissue to the prion surveillance center prior to performing any in-house processing of the tissue. Neurosurgery must speak with pathology prior to boarding the patient for surgery.

C. **For Autopsies:** The Anatomic Pathology departments in Corewell Health East will no longer perform autopsy services on deceased patients clinically suspected of harboring prion proteins (i.e. Creutzfeldt-Jakob Disease and other prion diseases). If there is any clinical suspicion of prion disease in the deceased patient, the Department of Decedent Affairs or appropriate authority (such as nursing) will refer the next of kin to the National Prion Disease

Pathology Surveillance Center for the purpose of coordinating the transfer of the body to the nearest facility willing to perform the autopsy services.

III. DEFINITIONS:

A. **CJD** = Creutzfeldt-Jakob Disease = most common type of TSE found in humans

B. **Prion** = infectious, misshaped protein found in TSEs that causes the deterioration of brain tissue and the infectivity of TSEs

C. **TSE** = transmissible spongiform encephalopathy = group of fatal, untreatable rapid dementia caused by prions, causing deterioration of mental and physical abilities, with lots of tiny holes being seen microscopically in the brain, similar to a sponge (= spongiform).

IV. COMMUNICATION:

A. When a patient suspected of CJD is boarded for surgery, the Operating Room (OR) staff at that site are responsible for communicating with the local pathology laboratory.

B. When a suspected CJD specimen is being transported from a laboratory to the Royal Oak Surgical Pathology laboratory, an email must be sent to the following recipients at Royal Oak:
   - System Lead Pathologist Assistant
   - Histology Manager
   - Histology Supervisor

C. Royal Oak Surgical Pathology alerts one of the neuropathologist attendings that there is a suspected CJD case that is being assigned to them.

D. For Autopsy: When CJD is in the differential diagnosis for dementia in a deceased patient, the Department of Decedent Affairs or appropriate authority (such as nursing) will refer the next of kin to the National Prion Disease Pathology Surveillance Center for the purpose of coordinating the transfer of the body to the nearest facility willing to perform the autopsy services. Should the Department of Pathology be contacted for autopsy, they will convey this information to the caller. They will inform the caller that such autopsies are not being performed at Corewell Health Hospitals.

National Prion Disease Pathology Surveillance Center
Address: Institute of Pathology
2085 Adelbert Rd Rm 419
Case Western Reserve University
Cleveland, OH 44106-4907 USA
Phone: 216-368-0587
Fax: 216-368-4090
Email Address: cjdsurv@case.edu
Website: http://www.cjdsurveillance.com

V. SPECIMEN COLLECTION AND HANDLING:

A. At the time of surgical excision, tissue must be separated into two containers of 10% Neutral Buffered Formalin:

   1. In Container A – a minimum of 1cm^3 biopsy of cerebral cortex and white matter

should be submitted for shipment to the Prion Surveillance Center.

2. In Container B – a minimum of 0.5cm^3 biopsy of cerebral cortex and white matter to be held at Royal Oak for possible surgical pathology.
3. If only a single specimen is sent down to the laboratory, the pathology laboratory will be responsible for splitting the tissue prior to sending to Royal Oak. For specimens grossed in the pathology laboratory:
    a. **Safety Considerations:** Grossing should be performed in a chemical fume hood. Disposable and fluid resistant gowns, shoe covers/leggings, and hats must be worn while handling of the tissue in histology. Protective wear should be removed before leaving the area. Protective wear should be worn only once. Mask with safety glasses, safety goggles or face shields (visors) must be worn while doing the autopsy and subsequent handling of the tissue and must be removed before leaving the grossing area. Kevlar gloves should be worn under two (2) pair of nitrile gloves. Disposable scalpels, preferably with rounded tip should be used.
    b. **Splitting the tissue into two containers:** Set up a plastic-backed, fluid absorbing pad/mat to set the container of tissue and to gross. Use a disposable cutting board, or something disposable, such as a petri dish. Specimen manipulation should occur using disposable forceps and a disposable scalpel.
    c. **Material Disposal:**
        i. All disposable instruments (e.g., scalpel handles, blades, forceps.), are to be decontaminated.
            a. Soaked in 2% (20,000 ppm) bleach (2 parts bleach, 3 parts water) for one (1) hour
            b. Dispose in the sharps container.
        ii. All disposable items (e.g., towels, gauze, extra tissue pieces/ shaving, mats, gloves, gowns, goggles, etc.) are to be decontaminated.
            a. Soaked in concentrated 2% (20,000 ppm) bleach (2 parts bleach, 3 parts water) for one (1) hour
            b. Placed in a double biohazard bag
            c. Placed in a corrugated box
            d. Mark box for “incineration”
            e. Place near the red biohazard bins
            f. Fill out “Generator Certification of Pre-Treatment” form, available from SteriCycle or Corewell Health Safety Department
        iii. Surfaces: (e.g., floors, tables, counters, hood work surfaces, etc.), contaminated with high risk tissue, are to be decontaminated:

a. Remove excess tissue from surface, as excess organic material will reduce the strength of the bleach or sodium hydroxide

b. Soak with either 2% (20,000 ppm) bleach (2 parts bleach, 3 parts water) for 1 hour

c. Keep entire surface wet the entire time

d. Cover with plastic (such as plastic bag), taped down

e. Rinse well with water or 95% alcohol.

4. For cases done outside Royal Oak:
    a. On the outside of each specimen container a label should be placed including patient demographics and specimen source.
    b. Each specimen should be placed in a leak-proof polybag with the absorbent material and seal shut. The sealed polybag containing the sample should then be placed into a Tyvek envelope and sealed shut. On the outside of the envelope, the location to which the specimen should be sent (Prion Surveillance Center or Royal Oak Surgical Pathology) should be written.
    c. Send one of the two samples directly to the Prion Center and the other to Royal Oak Surgical Pathology with the required documentation.
5. For Royal Oak specimens:
    a. The neuropathologist will go to the OR to pick up the specimens. These specimens will be brought down to the laboratory for packaging for send out.

B. Transport to Royal Oak campus:
    1. If the biopsy is being performed at a site other than Royal Oak, the two specimen containers will be moved to Royal Oak – Anatomic Pathology with all required paperwork.
    2. Both Tyvek envelopes should be placed into a cooler with the following paperwork and zip-tied shut for the courier:
        a. Requisition/Operative Note
        b. Patient Demographic Face Sheet
        c. National Prion Disease Pathology Surveillance Center (NPDPSC) Test Request form and full family history

VI. SPECIAL SAFETY PRECAUTIONS:

Tissue is considered a high risk specimen for the potential of CJD transmission. Standard disinfectants including those that contain formaldehyde, alcohol or other commercial germicidal products are not effective at inactivating prions. Incineration, Sodium Hydroxide, Formic Acid and Bleach are effective at specific percentages, times and temperatures.

VII. PROCEDURE:

A. National Prion Disease Pathology Surveillance Center (NPDPSC) Test Request form:
    1. The local pathology laboratory completes the test request form including the neurologists contact information and the pathologists' contact information
    2. The most up to date version of the NPDPSC test request form can be found at: https://case.edu/medicine/pathology/divisions/prion-center/
    3. Select for sample enclose: Fixed brain biopsy tissue in 10% Neutral Buffered Formalin, input biopsy and select the box that states the specimen was not treated with Formic Acid
    4. Must include all required clinical history

B. Upon arrival in the laboratory at Royal Oak the specimen is accessioned into Beaker.

C. Histology transfers one of the two specimen containers to Corewell Health Laboratory Send-Outs Department to be sent to the NPDPSC. Specimen two is held in the histology fume hood.

D. Once the results are returned from the NPDPSC, the secondary specimen container held at Royal Oak is processed using one of two methods:
    1. If the NPDPSC calls the specimen negative, the residual tissue at Royal Oak is treated as a stat specimen to get the slides out to the neuropathologist assigned the case in a timely manner
    2. If the NPDPSC calls the specimen positive, the department pays the NPDPSC to make teaching slides to be used for the residency program. Container B is disposed of using the following procedure:
        a. Cross of the name on specimen container with a black marker
        b. Place into a large white bucket
        c. Using a marker, label bucket: "FOR INCINERATION – POSSIBLE CJD"
        d. Prepare for Stericycle incineration by performing the following:
            i. Placed bucket in a corrugated box
            ii. Mark box for "incineration"
            iii. Place near the red biohazard bins
            iv. Fill out "Generator Certification of Pre-Treatment" form, available from SteriCycle or Corewell Health Safety Department. Attach to box.

VIII. REFERENCES:

A. National Prion Disease Pathology Surveillance Center
B. Biosafety in Microbiological and Biomedical Laboratories—6th Edition (cdc.gov)

Approval Signatures

Step Description	Approver	Date
CLIA Site Licensed Medical Directors	Muhammad Arshad: Chief, Pathology	9/27/2024
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CLIA Site Licensed Medical Directors	Jeremy Powers: Chief, Pathology	9/12/2024
CLIA Site Licensed Medical Directors	Ryan Johnson: OUWB Clinical Faculty	9/11/2024
CLIA Site Licensed Medical Directors	Subhashree Mallika Krishnan: Staff Physician	9/10/2024
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CLIA Site Licensed Medical Directors	John Pui: Chief, Pathology	9/10/2024
CLIA Site Licensed Medical Directors	Masood Siddiqui: Staff Pathologist	9/10/2024
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Operations Directors	Elzbieta Wystepek: Dir, Lab Services	7/17/2024
Operations Directors	Christopher Ferguson: Dir, Lab Services	7/17/2024
	Christopher Ferguson: Dir, Lab Services	7/17/2024

Applicability

Dearborn, Farmington Hills, Grosse Pointe, Royal Oak, Taylor, Trenton, Troy, Wayne