

# Creutzfeldt-Jakob Disease A Practical Guide for the Embalmer

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## Introduction

Creutzfeldt-Jakob Disease (CJD) is caused by an agent that appears to be resistant to chemical and physical agents normally used for sterilization and decontamination. Therefore, the precautions and recommendations presented here must be considered only preliminary.

The challenges presented here will magnify as environmental issues relating to medical waste increase. Although the risk of CJD transmission is low, it does exist and therefore must be addressed. Effective measures of prevention must be developed and tested, with recommendations derived from research specific to the handling and preparation of infected deceased remains.

Current recommendations have been made on the basis of the available data. As our knowledge of CJD increases, our recommendations may require revision.

## CJD Defined

Creutzfeldt-Jakob disease (CJD) is an infectious, progressive, degenerative neurological disease that can afflict anyone. There is no known treatment, and the disease is always fatal. The causative agent is extremely hardy and resistant to all measures of decontamination and sterilization routinely used in funeral homes. It can live for long periods of time in a dried state and challenges the routine decontamination measures practiced today.<sup>1</sup>

## Recommendations for Funeral Directors

### Transfers/Removals

The staff member making the transfer of the deceased from the place of death to the funeral home should follow normal removal personal protection procedures.

### Embalming Non-Posted Remains

1. *Protective Attire.* Prior to preparation the embalmer and assistant should follow universal precautions (as advocated for all other embalmings). Protective attire should include a disposable fluid resistant coverall or gown and pants, a cover apron, heavy latex gloves or double latex surgical gloves (note: vinyl gloves do NOT provide adequate barrier protection)<sup>2</sup>, a disposable surgical mask, hair cover and shoe covers and suitable eye protection; goggles or full face splash shield.
2. *Positioning.* The remains should be placed on the embalming table and post mortem wrappings removed, along with clothing, personal effects, and valuables. Any wrappings or clothing soiled with bodily fluids should not be reused and should be disposed of properly.
3. *Topical Disinfection.* The entire body, including orifices, are to be topographically disinfected with chemicals appropriate to the task or

## **Embalming Non-Posted Remains, continued**

with diluted arterial fluid (concentrated embalming fluid mixed with water).

4. *Washing.* Facial hair, if present, is to be removed by shaving. The entire body, including hair, should then be washed with a solution of germicidal soap.
5. *Manipulation.* Rigor mortis may be relieved by flexing the limbs through the application of massage and manipulation. The remains should be straightened and placed in a relaxed position with the head elevated above the chest. The arms may be placed at the decedent's side or folded on the abdomen. The mouth and eyes should be closed.
6. *Injection Site Selection.* The embalmer may select the appropriate vessels for injection of the preservative fluid and drainage of blood. It is important to recognize that the ability to transmit CJD via blood remains unproved. Additionally most body secretions such as urine, feces, milk, saliva and semen are considered non-infectious. The major concern to the embalmer is contact with the brain, the spine and cerebral spinal fluid, which should not pose a significant concern embalming a straight "non-autopsied" case
7. *Injection Chemicals.* The embalmer may determine the appropriate selection, mixture and dilution of

embalming chemical to be injected as usual. Preservative fluid(s) are diluted with water in the embalming machine in an average quantity of one to four gallons total dilute mixture. In that formaldehyde, glutaraldehyde, formalin and phenols are all ineffective or unreliable methods for the eradication of CJD prions, no special fluids are suggested at this time. The purpose of arterial injection should be considered for preservation and cosmetic purposes only, NOT for disinfection or eradication.

8. *Arterial Injection.*
  - a. The dilute chemical solution may be injected into the raised artery through the use of varying rates of pressure, forcing blood to drain out of the accompanying raised vein via a drain tube and an appropriate length hose. The embalmer may analyze fluid distribution throughout all areas of the deceased to ensure proper distribution and preservation.
  - b. Secondary and subsequent points of injection and drainage may be made, should areas require additional injection and preservation. When the embalmer is comfortable with the level and quality of preservation, the injection process may conclude.
  - c. Injection tubes are removed, the areas of injection are dried, tightly

**Embalming Non-Posted Remains, continued**

sealed with absorption chemicals and/or adhesive chemicals, and sutured.

**9. CAUTION: No Cavity Aspiration.**

Under NO circumstances should aspiration of the abdominal and thoracic cavities take place. Certain viscera (liver, lung, kidney & spleen) have transmitted this infection, although with less predictability than direct contact with cerebral spinal fluid. Given the possibility that minute tissue may become part of the aspirate the remains should NOT undergo aspiration or cavity injection.

**10. Aspiration Alternative.** Instead of cavity aspiration, the embalmer should carefully introduce eight ounces of cavity fluid intraorally, massaging the throat area externally so that the fluid is introduced internally via the esophagus and bronchi. Attaching the cavity injector to the nasal aspirator as a controlled method of delivery works well in these instances and reduces the potential of cavity fluid coming into contact with mucous membranes. This will slow down bacterial migration from the respiratory and upper gastrointestinal tract.

**11. Additional Precautions**

a. The embalmer should also pack the nose, mouth, and all other orifices

with treated cotton. This will help to reduce visceral decomposition, bloating and purge.

b. Should purge be a concern of the embalmer, minimal pressure should be utilized for injection, a nasal aspirator may be utilized in the nose, throat and mouth, and the nose and throat should be appropriately packed with cotton or Webril, pretreated with a phenol-based solution to prevent wicking and to create a chemical barrier between the purge and the external environment.

c. The thoracic and abdominal cavities should not be directly injected with undiluted preservative fluid (cavity fluid) nor should the embalmer use a trocar for any purpose.

d. Any remaining surgical device (e.g., intravenous tubes), wounds or openings should be cauterized with an appropriate phenol-based chemical, sutured with ligature, and sealed.

**12. Washing.** The body is washed for the second and final time with dilute bleach, rinsing with water. The remains, including hair, may be washed with germicidal soap or shampoo.

**13. Visitation.** Remains prepared in this fashion are suitable for visitation.

**14. Instruments.** By following the above guidelines, the instruments used in a

## **Embalming Non-Posted Remains, continued**

non-posted embalming would be considered low-risk and may be cleaned and disinfected or sterilized using conventional protocols of heat or chemical sterilization or high level disinfection.

Although the use of disposable instruments is preferred, steam sterilization is recommended for the processing of re-usables; 18 minutes at 270° F pre-vacuum, or 60 minutes at 270° F gravity cycle.

Unfortunately many preparation rooms do not contain steam sterilizers, thus requiring the use of chemicals for instrument disinfection in either a 1:10 dilution of household bleach for one hour contact time or a conventional phenolic disinfectant for one hour.

15. *Surfaces.* Floors, walls, countertops or other environmental surfaces should be cleaned with a low-level disinfectant in the conventional fashion. A 1:10 dilution of household bleach can be used to spot decontaminate visible residues of tissue before cleaning.
16. *Disposables.* All disposables and/or contaminated waste should be properly disposed of as a “sharp” or “medical waste.”

## **Further Recommendations, Embalming Posted Remains**

Note: See “Embalming Non-Posted Remains” for other important information.

In a post mortem case, the remains have been autopsied and any or all of the cranial, abdominal and thoracic cavities may have been eviscerated. According to the College of American Pathologists, Guidelines for High Risk Autopsy Cases, in the future, as in the past, it may be desirable for purposes of medical research to conduct complete autopsies on CJD patients.<sup>3</sup>

We recommend the following procedures for embalming an “autopsied” CJD case:

1. Should viscera be present, it should remain within the bag provided.
2. Preservative powder/fluid/gel should be added into the viscera bag and the bag should be closed and placed within a second bag.
3. Upon completion of injection all liquid should be aspirated from the thoracic and abdominal cavities and the internal surfaces treated with a preservative gel or powder.
4. The viscera bag should be placed and sutured within the thoracic and abdominal cavities.
5. The cranial cavity should be dried and the walls treated with a preservative gel or powder.
6. The calvarium may now be re-attached and the incision sutured.

## Decontamination

A serious epidemiological concern in funeral service is how to destroy the infectious agent, which is extremely hardy and resistant to heat, formaldehyde, formalin, phenol, glutaraldehyde, ionizing radiation, freezing, drying and organic detergents.<sup>4</sup> Based on the epidemiology of iatrogenic and nosocomial episodes of CJD, it is clear that the only exposures in patient care settings which have resulted in infection are those instances involving devices which cannot be cleaned and which are contaminated with high-risk tissue from the central nervous system (e.g., brain, cerebrospinal fluid, corneas), or, in the case of transplants, exposures due to direct and intimate contact with CJD-laden brain tissue.

### Current Findings

As previously stated, CJD has been transmitted via contaminated surgical instruments. When a device is contaminated with tissues or body fluids that are not high risk, and the device is cleanable, the probability of infection transmission appears to be so low that it would not be measurable.

Based on studies of the agent that causes scrapie, alcohol, boiling, detergents, dry heat, ethylene oxide, hydrogen peroxide, iodophers, ionizing/ultraviolet radiation, peracetic acid, and steam sterilization are all ineffective or unreliable methods for the

eradication of prions, as are formaldehyde, formalin, glutaraldehyde and phenols, the primary ingredients in modern embalming chemicals.

1. *Formaldehyde and Glutaraldehyde.* Both glutaraldehyde and formaldehyde are fixative chemicals and would render prion/tissue more stable. Although extensive studies on other generic chemical germicides have not been done, it has been shown that a conventional phenolic disinfectant was effective in reducing the prion load by up to 7 logs.

Formaldehyde renders the organism virtually indestructible, even when subjected to ashing (the application of dry heat at 360 degrees C for one hour).

Immersion in glutaraldehyde for three weeks is only partially effective in decontaminating CJD. Steam sterilization at 134 degrees C produced inconsistent results.

2. *Bleach.* Sodium hypochlorite (bleach) is recommended as a decontaminant for CJD in a funeral home setting although its effectiveness as a treatment has been inconsistent. The chemical is extremely corrosive to metal, especially stainless steel, damaging instruments and preparation room equipment, and the chlorine fumes irritate the respiratory tract.<sup>5, 6</sup>

## Decontamination continued

3. *Lye.* Inconsistent results were found in residual prion infectivity after contact with sodium hydroxide (lye). Although sodium hydroxide is less caustic than hypochlorite, it corrodes aluminum and is a hazardous substance requiring neutralization prior to disposal. Additionally, it deteriorates significantly within several months of storage.<sup>6, 7</sup>
  4. *Peroxide and Peracetic Acid.* Neither hydrogen peroxide nor peracetic acid are able to eradicate prions.<sup>8</sup>
  5. *Standard gravity sterilization.* Standard gravity sterilization at 121 degrees C for up to 120 minutes is unreliable, as is dry heat at 160 degrees C for 24 hours.
- However, standard gravity sterilization at 132 degrees C (270 degrees F) for 60 minutes effectively eradicated CJD and scrapie infectivity in intact brain tissue.
6. *Incineration.* Although incineration destroys prions and is consistently recommended in healthcare settings, no funeral home is equipped to incinerate on site as in a hospital setting. Additionally, new DEP regulations place such enormous regulations on existing hospital incinerators that a large percentage of existing incinerators are expected to be shut down.

With incineration the treatment of choice for all contaminated disposable items, proper containment and disposition of solid

medical waste should continue following normal operating procedures. Floors, walls, counter-tops, or other housekeeping surfaces in medical wards, autopsy rooms, laboratories and preparation rooms that are contaminated with patient tissues known to contain CJD should be cleaned with a suitable detergent in the conventional fashion. A 1:10 dilution of household bleach can be used to spot decontaminate visible residues of tissue before cleaning.

## CJD: What We Know Today

### Overview

The organism causing CJD is a transmissible agent—much smaller than a virus—that greatly resists chemical and physical agents. These organisms are proteineaceous and infectious, and they resist inactivation by most procedures that modify nucleic acids. The term “prion” (pronounced pree-on) was coined by Prusiner in 1982 to describe the agents responsible for a group of chronic progressive central nervous system (CNS) disorders that share similar pathologies.<sup>9, 10</sup>

Prions cause “spongiform encephalopathy” in animals and humans. Animal prion diseases include scrapie in sheep, chronic wasting disease in elk and deer, transmissible mink encephalopathy, feline spongiform encephalopathy in cats and bovine spongiform encephalopathy (BSE or Mad Cow Disease, which displays

similarities to CJD) in cows. In humans, prions cause CJD, fatal familial insomnia, Gertsmann-Straussler-Scheiner Disease, and Kuru, a neurologic disease similar to CJD and found only among a few primitive tribes in the eastern Papua regions of New Guinea. The decline in Kuru has been attributed to the cessation of ritual cannibalism, the consumption of dead kinsmen that had been practiced as a ritual of mourning. For years, women and children of the Fore tribe who ate the brains and other organs of deceased relatives in solemn ceremonies became infected with the disease. This disease became known as “Kuru,” which means trembling or fear. Kuru was the first chronic “degenerative” CNS disease of humans shown to have a slow transmission.

### **Diagnosis**

CJD was first identified simultaneously by Creutzfeldt and Jakob in the early 1920's.<sup>11</sup> Although Jakob initially suggested that the disease might be infectious, this was not confirmed until more than 40 years later.

Definitive diagnosis is made by a brain biopsy obtained surgically or on postmortem examination. The infected brain tissue contains vacuoles, which give the tissue a “spongelike” appearance on a microscopic level. For this reason, CJD and related disorders are called transmissible spongiform encephalopathies.<sup>12</sup>

In patients with dementia, the location of a specific brain protein in the cerebrospinal fluid strongly supports a diagnosis of CJD. This new finding, however, does not support the use of this test in patients without clinically evident dementia.<sup>13</sup>

### **Transmission**

Three forms of CJD have been described: genetic, from human to human, which accounts for 10 - 15% of all CJD cases and is caused by mutations within familial gene encoding; iatrogenic or infectious, where the prion is introduced from an external source, such as corneal transplants or dura mater grafts, or from a therapeutic misadventure (medical or surgical treatment that results in an unfavorable response); and sporadic, which accounts for the majority of cases. Sporadic CJD is a result of either mutations arising in the tissue and accumulating over time, or from an infectious transmission from an unknown source.<sup>14</sup>

The natural transmission of CJD is not understood, primarily because of difficulties determining causality after a long incubation period. The incidence rate is less than one case per million people per year.<sup>15</sup> CJD has been found throughout the world, with a prevalence reported in large population centers where the disease is more readily diagnosed.<sup>16</sup>

Skin and most bodily secretions and excretions (e.g. urine, feces, milk, saliva,

## **Transmission continued**

semen) are considered noninfectious.<sup>17</sup> Transmissions have occurred via transplanted tissue; cadaver extracted hormones used as a growth treatment for dwarfism and short stature; and through the use of contaminated surgical instruments. Transplanted tissue including cornea, pericardial homograft and dura mater has transmitted the disease to recipients.<sup>18</sup> The risk of CJD transmission is a function of the type of tissue and the expected relative concentration of its agent; the risk is high for contact with brain and corneal tissue, medium for cerebrospinal fluid, kidney, liver, lymph nodes, and spleen tissue, and low to none for blood, urine, feces, nasal mucus, saliva, sputum, tears, heart, adrenal, bone marrow, muscle, and nerve tissue.

The exact mode of transmission in humans is not known. Transmission studies have shown that primates can be infected via percutaneous inoculation but not by simple direct contact. Transmission of CJD has not been associated with environmental contamination. Brain, spinal cord and cerebrospinal fluid from humans or animals with CJD have regularly transmitted infection when inoculated into animals. Other viscera (liver, lung, kidney, spleen) transmit the infection with less predictability.

The ability to transmit CJD via blood transfusions is only speculative and

remains unproved. Blood has been found to be ineffective as a medium of CJD transmission in current studies using guinea pigs.<sup>19</sup> For embalmers, this would suggest that the preparation of a body infected with CJD presents no more risk than a body with any other disease.

Isolated episodes of CJD have occurred in approximately 24 physicians and other health care workers including two neurosurgeons, one pathologist, nine nurses, and two histology technicians. The incidence of CJD in these groups does not exceed what would be expected by chance alone. Throughout the world there have been no documented reports of transmission of disease from patients to hospital or mortuary staff.<sup>20, 21</sup> However, the unusual resistance of the agent to inactivation necessitates special precautions in dealing with infected individuals.

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