

Human Prion Diseases

Recommendations for Funeral and Crematory Practitioners

Background

Prion diseases, also referred to as transmissible spongiform encephalopathies (TSE), are rare and fatal neurodegenerative diseases of animals and humans. “Prion” stands for *proteinaceous infectious particles*. The causative agent of prion diseases is thought to be a misfolded infectious isoform, called PrP^{Sc}, of a normally occurring cellular protein, PrP^C. This abnormal folding process can occur spontaneously (sporadic), by genetic mutations (familial), or by the uptake of prions from an external source (iatrogenic, variant). The resulting accumulation of abnormal protein in the central nervous system causes progressive neurodegenerative spongiform changes.

Creutzfeldt-Jakob disease (CJD) is the most common human prion disease with an incidence of 1-2 cases per million population per year. Sporadic CJD (sCJD) occurs for unknown reasons and accounts for approximately 85-90% of cases. Familial CJD (fCJD) results from an inherited mutation in the prion protein gene and accounts for about 10-15% of cases. Less common inherited forms include Fatal Familial Insomnia (FFI) and Gerstmann-Straussler-Scheinker Syndrome (GSS).

Historically, iatrogenic cases (iCJD) have been associated with human-derived pituitary hormone, dura mater grafts, corneal grafts, and contaminated neurosurgical equipment. All of the equipment-related cases occurred before routine implementation of the sterilization procedures currently used in health care facilities. No equipment-related cases have been reported since 1976. In the United States, 29 iCJD cases have been linked to the use of pituitary human growth hormone (hGH) in patients treated before 1977. The growth hormone now used for treatment poses no threat of infection with CJD.

Variant CJD (vCJD) is associated with consumption of cattle products contaminated with the prion agent causing bovine spongiform encephalopathy (“mad cow disease”). To date, no case of variant CJD acquired in the United States has been documented.

Misconceptions

Unlike airborne transmitted illnesses such as tuberculosis and influenza or those transmitted by blood or sexual contact such as hepatitis and HIV, CJD is not transmissible person-to-person. Many funeral and crematory practitioners encounter infectious diseases and are able to serve families and protect the health of the public. CJD should not be an exception. In fact, it is less infective compared with many common infectious diseases and does not present a substantial risk to the health and safety of others.

Handling of bodies of CJD patients

Transporting

- Funeral service staff can safely remove the body from a hospital ward, hospice, or other setting and transport it to a funeral home using standard infection control measures.
- Prior to moving the body, place it in a leak-proof bag lined with absorbent material.
- If national or international transportation is needed, it will be necessary to comply with IATA (International Air Transport Association) and ICAO (International Civil Aviation Organization) Restricted Articles Regulations along with any additional requirements of the specific carrier. It should be noted that IATA requires embalming of the body.

Preparation/Embalming

Unautopsied bodies

- Routine contact or handling of an intact body does not pose a risk. Cosmetic work can be done using standard precautions.

- Avoid surface contamination from perfusion drain sites by placing the body on an impermeable sheet.
- All drainage fluids should be collected into stainless steel containers.
- Perfusion sites should be closed with cyanoacrylates and wiped with bleach.

Autopsied bodies

- Embalming an autopsied body is not encouraged, but can be safely done.
- Disposable masks, gowns, and puncture resistant gloves are strongly recommended.
- If possible, use disposable instruments.
- Use a plastic sheet with absorbent wadding and raised edges underneath the head.
- Avoid surface contamination and contain suture site leakage by placing the body on an impermeable sheet or body pouch.
- All drainage fluids should be collected into stainless steel containers.
- If sutures do not completely control leaking, the cranial cavity should be packed with absorbent material that has been soaked with bleach then tightly sutured.
- If viscera are present, they should remain within the bag provided.
- Add preservative powder/fluid/gel to the viscera bag, close it, and place it in a second bag.
- The cranial cavity should be dried and the walls treated with a preservative gel or powder.
- Perfusion and autopsy incision sites should be closed with cyanoacrylates.
- The entire body should be wiped down with bleach, with special attention to any autopsy incision and perfusion sites.

Disinfection and waste removal instructions are the same for both autopsied and unautopsied bodies. (See below)

Casketing

- Avoid unnecessary manipulation that might force purging of body fluids and risk opening incision sites.
- CDC recommends that, for bodies that have been autopsied, family members should avoid superficial contact (e.g. touching or kissing the face).
- For patients that have not been autopsied, contact need not be discouraged.

Cremation and Burial

- No special interment, entombment, cremation, or inurnment requirements are needed.
- Interment of closed caskets do not present a significant risk of environmental contamination.
- Cremated remains are considered sterile. The CJD agent does not survive incineration temperatures.

Disinfection and Waste Removal Instructions

Solid Materials

- All plastic sheets and other disposable items exposed to bodily fluids should be incinerated.
- Contaminated solid materials should be disposed of as hazardous waste.
- Reusable instruments can be cleaned and disinfected by using CDC recommendations available at <https://www.cdc.gov/prions/cjd/infection-control.html>

Collected fluids

- Add 40 grams of sodium hydroxide pellets per liter of collected fluid.
- Stir the mixture for a few minutes, avoiding any spilling (the fluid will be hot).
- Leave the mixture undisturbed for at least one hour, then dispose of it like other mortuary waste.

Working Surfaces

- Surfaces that have been contaminated should be flooded with sodium hydroxide or bleach and left undisturbed for at least one hour.
- Using gloves, mop up with absorbent disposable rags and then swab with water sufficient to remove any residual disinfectant solution.

Body fluids, tissues, and hazardous chemicals should be handled in accordance with routine funeral home policies, and state and federal regulations.

Resources

CDC Information for Funeral and Crematory Practitioners

<https://www.cdc.gov/prions/cjd/funeral-directors.html>

WHO Infection Control Guidelines for Transmissible Spongiform Encephalopathies. See Section 8.

<http://www.who.int/csr/resources/publications/bse/whocdscsraph2003.pdf?ua=1>

If you are a funeral director or embalmer and have further questions regarding handling a body affected by CJD, please contact the National Prion Disease Pathology Surveillance Center for guidance to relevant information or for further assistance: (216) 368-0587 or CJDsurveillance@uhhospitals.org

The [CJD Foundation](#) can provide case-by-case advice and guidance through their HelpLine: 1 800 659-1991

Contact Information

- Local health departments:
<https://www.doh.wa.gov/AboutUs/PublicHealthSystem/LocalHealthJurisdictions>
- Washington State Department of Health, Office of Communicable Disease Epidemiology:
(206) 418-5500 or (877) 539-4344 or
<https://www.doh.wa.gov/AboutUs/ProgramsandServices/DiseaseControlandHealthStatistics/CommunicableDiseaseEpidemiology>