Michigan Department Of Community Health

MDCH

Creutzfeldt-Jakob Disease (CJD) Fact Sheet for Funeral Directors

Adapted from the New York State Department of Health, "Information on Creutzfeldt-Jakob Disease For Funeral Home, Cemetery, And Crematory Practitioners"

What is CJD?

CJD is a rapidly progressive and fatal disease caused by the accumulation in the brain of an abnormal form of a protein known as a "prion." CJD affects approximately one person per million population each year. The disease has a very long incubation period, ranging from 15 months to 30 years. The average age of CJD symptom onset is around age 60. Initial symptoms of CJD generally include progressive dementia, confusion, behavioral changes, and muscle incoordination. Other symptoms may include depression, insomnia, and problems with vision and speech. The progression of the disease is rapid and nearly all individuals diagnosed with CJD die within one year.

How is CJD Diagnosed?

The use of clinical signs and various medical tests, such as CT scans and MRIs, can provide support for the diagnosis of CJD. The only way to confirm a diagnosis of CJD, however, is from a brain biopsy or autopsy. A direct examination of brain tissue is needed in order to determine the presence and type of prion disease.

What are the different types of CJD?

Sporadic CJD- accounts for approximately 85-90% of CJD cases. The disease results from the spontaneous conversion of the normal brain protein into the disease-causing abnormal prion protein.

Familial CJD- is an inherited form of CJD caused by a genetic mutation and accounts for 5-15% of cases. This gene mutation makes it more likely that the brain protein will convert to the abnormal disease-causing form.

Iatrogenic CJD- is transmitted by direct exposure to abnormal prion proteins from an external source during a medical procedure. In rare situations, CJD has been spread by the re-use of contaminated surgical instruments or the transplantation of certain high-risk tissues from a CJD infected donor.

Variant CJD (vCJD)- is the human form of bovine spongiform encephalopathy (BSE) or "Mad Cow Disease." Variant CJD is linked to the consumption of meat from an animal infected with BSE or by blood transfusion from a donor infected with variant CJD. Variant CJD is caused by a different prion protein from the sporadic or familial forms of CJD. To date, no cases of variant CJD acquired in the U.S. have been found.

Is CJD contagious?

CJD is not contagious in the typical sense, and is not transmitted person to person by direct contact, airborne spread, or the environment. CJD transmission can occur during invasive medical procedures involving the central nervous system due to exposure to contaminated brain tissue. This accounts for less than one percent of all CJD cases. Standard disinfectants and routine embalming solutions are ineffective against "prions;" chemical solutions and physical processes involving bleach, sodium hydroxide, or autoclaving must be used to inactivate the prion.

How should the body of a person with a prion disease be transported?

Mortuary workers can safely remove the body of a CJD patient from the place of death and transport it to the funeral home preparation room using appropriate standard infection control measures, which includes wearing personal protective gear. The World Health Organization (WHO) recommends placing the body in a leak-proof pouch prior to moving. The bag should be lined with absorbent material to prevent leakage of body fluids. In instances where there is excess fluid, a double bag can be utilized. After transporting, all surfaces (i.e., stretchers, cots) should be disinfected with bleach.

How should the body of a person with a prion disease be prepared?

Embalming bodies of CJD patients who have **not been autopsied** can be performed using standard precautions. It is suggested that the body be placed on a waterproof sheet to collect bodily fluids and that disposable instruments be used. The bodily fluids should be collected in a suitable container. Incision sites should be closed with super glue, wiped down with bleach and the body washed prior to dressing. Cosmetic restorative work may also be completed.

Embalming bodies of CJD patients **who have been autopsied** can also be safely performed. Adherence to standard infection control measures is very important when embalming an autopsied body of a suspected or clinically diagnosed CJD patient. Autopsies on these individuals are often restricted to removal of the brain, therefore, special precautions should be taken including placing a plastic sheet with absorbent wadding and raised edges underneath the head to ensure containment of fluids and prevent any spillage. In instances where sutures do not completely control leaking, the cranial cavity should be packed with absorbent material that has been soaked with bleach, and tightly sutured.

Bodies of **autopsied CJD patients** should be placed on a waterproof sheet to collect all fluids. It is strongly recommended that disposable instruments, masks, gowns, and puncture-resistant gloves (vinyl gloves do not provide adequate protection) be used whenever possible. The entire body should be washed with bleach, rinsed, and sanitized before dressing. Special care should be taken to limit fluid leakage when performing restorative work. All fluids should be collected in a suitable container.

How should the casketing and viewing be handled?

Unnecessary manipulation of the body that would force leaking of body fluids and risk opening of incision sites should be avoided. If needed, the casket can be lined with a leak-proof sheet. An open casket for viewing should not be prohibited. If an **autopsy has been performed,** family members of CJD patients should be advised to avoid superficial contact (such as touching or kissing the patient's face) with the body. However, if the patient has not been autopsied, such contact need not be discouraged.

What are the proper procedures for disinfection and waste removal?

According to the World Health Organization (WHO) Infection Control Guidelines for Transmissible Spongiform Encephalopathies, all collected fluids should be disinfected by adding 40 grams of sodium hydroxide pellets per liter of collected fluid. The mixture should be stirred after a few minutes and care should be taken to avoid spillage, as the fluid will be hot. It should then be left undisturbed for at least one hour, after which it can be disposed of like other mortuary waste. Plastic sheets and other disposable items that have been exposed to bodily fluids should be incinerated. Mortuary working surfaces that have accidentally become contaminated should be flooded with sodium hydroxide or bleach, left undisturbed for at least one hour, then (using gloves) mopped up with absorbent disposable rags, and surface swabbed with water sufficient to remove any residual disinfectant solution.

Work surfaces can be disinfected by flooding with undiluted bleach. Although the use of disposable instruments is preferred, reusable instruments and tools can be cleaned and disinfected by using CJD sterilization protocols recommended by the Centers for Disease Control and Prevention. All contaminated solid materials should be disposed of as hazardous waste. Disposing of body fluids and tissues and of hazardous chemicals should be handled in accordance with funeral home policy, state, and federal regulations.

What are the instrument sterilization methods in the WHO TSE infection control guidelines recommended by the CDC?

The methods below are listed in order of more (1) to less (3) stringent. Instruments should be decontaminated by a combination of the chemical and recommended autoclaving methods before subjecting them to cleaning in a washer cycle and routine sterilization. Washers should be run through an empty cycle after the processing of these instruments prior to any other routine use. Sodium hypochlorite may be corrosive to some instruments. The instrument manufacturer should be consulted about the instrument's tolerance of exposure to sodium hypochlorite prior to using these methods. FDA investigations suggest that much of the damage from autoclaving in sodium hydroxide is cosmetic and should not affect the performance or cleaning of the instruments.

1. Immerse instruments in a pan containing 1N sodium hydroxide (NaOH) and heat in a gravity displacement autoclave at 121°C for 30 min, clean, rinse in water, and subject to routine sterilization. [CDC NOTE: The pan containing sodium hydroxide should be covered, and care should be taken to avoid sodium hydroxide spills in the autoclave. To avoid autoclave exposure to gaseous sodium hydroxide, the use of containers with a rim and lid designed for condensation to collect and drip back into the pan is recommended. Persons who use this procedure should be cautious in handling hot sodium hydroxide solution (post-autoclave) and in avoiding potential exposure to gaseous sodium hydroxide. Exercise caution during all sterilization steps, and allow the autoclave, instruments, and solutions to cool down before removal.

Or

2. Immerse instruments in 1N NaOH or sodium hypochlorite (20,000 ppm available chlorine) for 1 hour; transfer instruments to water; heat in a gravity displacement autoclave at 121°C for 1 hour; clean; and subject to routine sterilization.

Or

3. Immerse instruments in 1N NaOH or sodium hypochlorite (20,000 ppm available chlorine) for 1 hour; remove and rinse in water, and then transfer to open pan and heat in a gravity displacement (121°C) or porous load (134°C) autoclave for 1 hour; clean; and subject to routine sterilization.

Are any special burial or cremation procedures needed?

No special interment, entombment, inurnment, or cremation procedures are required for deceased patients with CJD. Interment of bodies in closed caskets does not present a significant risk of environmental contamination. Cremated remains can be considered sterile, as the infectious agent does not survive incineration-range temperatures (1000° C).

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