Michigan Department Of Community Health

MDCH

Creutzfeldt-Jakob Disease (CJD) Fact Sheet for Infection Control Professionals

What is CJD?

CJD is a rare, rapidly progressive, and fatal neurodegenerative disease caused by an abnormal form of the brain prion protein. CJD 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 including ataxia and myoclonus. Other symptoms may include depression, insomnia, and problems with vision and speech. The progression of the disease is rapid; nearly all individuals diagnosed with CJD die within one year.

What are the types of CJD?

There are four distinct types of CJD.

1) Sporadic CJD is the most common type (~85-90% of cases) and is believed to be caused by the spontaneous conversion of the normal prion protein into the disease-causing abnormal form.

2) Familial CJD (~5-15% of cases) is an inherited form of CJD caused by a genetic mutation.

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

4) 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 infectious tissue (cornea, dura mater grafts, human growth hormone). No equipment-related cases have been reported for over 30 years, since the implementation of current, routinely used sterilization and disinfection procedures.

How is CJD Diagnosed?

The use of CT scans, MRIs, EEGs, clinical signs, and the use of a specific tests for elevated 14-3-3 or tau proteins in cerebrospinal fluid can provide supporting evidence for the diagnosis of CJD. The 14-3-3 and tau tests alone are not considered diagnostic; the only way a confirmed diagnosis of CJD can be made is by examination of brain tissue from a biopsy or autopsy. Prion disease testing including 14-3-3 and tau protein tests, brain biopsy and autopsy examination are available through the National Prion Disease Pathology Surveillance Center (NPDPSC) at Case Western Reserve University. For help with the coordination of an autopsy or other testing protocols, please contact the NPDPSC at (216) 368-0587. Information can also be found on the center's website, http://www.cjdsurveillance.com.

What special precautions need to be taken when caring for a person with suspected or confirmed prion disease?

CJD is not contagious in the typical sense, and is not transmitted by direct patient contact or airborne spread. Health care workers should use standard precautions when caring for a patient with confirmed or suspected CJD. Gloves should be used when coming into contact with blood or bodily fluids. Additional protective equipment such as gowns, masks, and eye protection should be used when the potential for mucous membrane exposure to infectious materials exists during a procedure. As in all healthcare situations, care should be taken when handling sharps to prevent self-injury with a used instrument.

What tissues and body fluids are considered "high-risk" for CJD?

High levels of infectivity are found most often in the central nervous system (CNS) tissues, including the brain, spinal cord and eye. These tissues should be considered "high-risk" and care should be taken to avoid occupational exposure to them. Infectivity is found at lower levels in the cerebrospinal fluid (CSF), lung, liver, kidney, lymph nodes, spleen, tonsils, and placenta. These tissues/fluids would be considered "low-risk" for infection. Infectivity has not been detected in a number of tissues and fluids by currently available testing. These include heart, skeletal muscle, peripheral nerve, adipose tissue, gingival tissue, adrenal gland, thyroid, prostate, testis, urine, feces, saliva, mucous, semen, milk, tears, and sweat. The infectivity status of blood has not been definitively classified. While no cases of transfusion-related sporadic CJD have ever been documented, 3 cases of probable variant CJD have been linked to blood donations from asymptomatic variant CJD infected donors. For more complete information on tissue infectivity please refer to the 2006 WHO Guidelines on Tissue Infectivity Distribution in Transmissible Spongiform Encephalopathies, http://www.who.int/bloodproducts/TSEPUBLISHEDREPORT.pdf

How should an occupational exposure to “high or low-risk” tissues or fluids be handled?

To date, no known cases of human prion disease have occurred due to an occupational exposure. While no prion-specific post exposure guidance exists, the World Health Organization (WHO) recommends a basic disinfection approach when dealing with an occupational exposure. Contamination of intact skin with fluid or tissue should be washed with soap (avoid scrubbing) and warm water. For needle sticks or lacerations, gentle bleeding should be encouraged followed by washing with soap (avoid scrubbing) and warm water. Splashes onto a mucous membrane should be flushed repeatedly with saline or tap water. All occupational exposures should be reported to the appropriate health and safety authorities at your facility.

How should medical instruments be cleaned?

Whenever possible, medical instruments and materials used on suspect or confirmed cases of CJD should be disposable or single-use and be destroyed by incineration after use. For contact with high infectivity tissues where disposable instruments are not available, the highest level of safety is attained by destroying the reusable instrument by incineration. When the destruction of reusable instruments is limited by cost or practicality, disinfection procedures developed by the WHO should be used for decontamination of instruments that come into contact with high and low infectivity tissues from a known or suspected case of CJD. The CDC recommends the use of one of the three most stringent disinfection methods listed below. These stringent sterilization methods should also be used to reprocess medical instruments that come in contact with high infectivity tissues of persons known to be blood relatives of patients with familial forms of prion disease. Reusable instruments should be kept moist to prevent the drying of tissue and fluids on the item and cleaned as soon as possible. Instruments used on high and low infectivity tissues should be kept separate from those used on tissue where infectivity has not been detected. All used instruments should be contained in a leak and puncture-proof container labeled “biohazard” when being sent for incineration or sterilization.

What are the 3 most stringent sterilization methods in the WHO Transmissible Spongiform Encephalopathy 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.

How should neurosurgical instruments be reprocessed when used on patients with no definitive diagnosis at the time of a procedure?

For patients, in whom the clinical diagnosis leading to the neurosurgical procedure remains unclear, the instruments should be reprocessed in the same manner as that for instruments used in procedures involving suspected or confirmed CJD patients. In some patients undergoing neurosurgery, a CJD diagnosis that is not suspected before the procedure may be confirmed after the neurosurgery. Unless a clear non-CJD diagnosis is established, these patients should be considered as potentially suspected CJD patients for all other infection control requirements. To prevent the destruction of non-disposable instruments from cases where the diagnosis is unknown, instruments may be quarantined until a diagnosis is confirmed. This should only be done if there is assurance that the instruments will have no way of being accidentally reintroduced into the circulating instrument population. If prion disease is ruled out as a diagnosis the instruments may be returned to circulation after proper sterilization.

How should heat-sensitive instruments or surfaces that come in contact with suspected or confirmed CJD patients be decontaminated?

Whenever possible, work surfaces that may come into contact with high or low infectivity tissues should be covered with disposable materials that can be incinerated after use. Surfaces and heat-sensitive re-usable instruments that come in contact with high infectivity and low infectivity tissues should be decontaminated by flooding with or soaking in 2N NaOH or undiluted sodium hypochlorite for 1 hour and rinsed repeatedly with water. Sodium hypochlorite may be corrosive to some instruments.

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