Questions and Answers: Creutzfeldt-Jakob Disease Infection-Control Practices

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What is Creutzfeldt-Jakob disease?

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. CJD occurs worldwide and the estimated annual incidence in many countries, including the United States, has been reported to be about one case per million population.

The vast majority of CJD patients usually die within 1 year of illness onset. CJD is classified as a transmissible spongiform encephalopathy (TSE) along with other prion diseases that occur in humans and animals. In about 85% of patients, CJD occurs as a sporadic disease with no recognizable pattern of transmission. A smaller proportion of patients (5 to 15%) develop CJD because of inherited mutations of the prion protein gene. These inherited forms include Gerstmann-Strawssler-Scheinker syndrome and fatal familial insomnia.

How is CJD diagnosed?

Physicians suspect a diagnosis of CJD on the basis of the typical signs and symptoms and progression of the disease. In most CJD patients, the presence of 14-3-3 protein in the cerebrospinal fluid and/or a typical electroencephalogram (EEG) pattern, both of which are believed to be diagnostic for CJD, have been reported. However, a confirmatory diagnosis of CJD requires neuropathologic and/or immunodiagnostic testing of brain tissue obtained either at biopsy or autopsy.

Have there been any reports of iatrogenic transmission of CJD?

Yes, iatrogenic transmission of the CJD agent has been reported in over 250 patients worldwide. These cases have been linked to the use of contaminated human growth hormone, dura mater and...
These cases have been linked to the use of contaminated human growth hormone, dura mater and corneal grafts, or neurosurgical equipment. Of the six cases linked to the use of contaminated equipment, four were associated with neurosurgical instruments, and two with stereotactic EEG depth electrodes.

All of these equipment-related cases occurred before the routine implementation of sterilization procedures currently used in health care facilities. No such cases have been reported since 1976, and no iatrogenic CJD cases associated with exposure to the CJD agent from surfaces such as floors, walls, or countertops have been identified.

How should surgical instruments used on suspected or confirmed CJD patients be reprocessed?

Inactivation studies have not rigorously evaluated the effectiveness of actual cleaning and reprocessing methods used in health care facilities. Recommendations to reprocess instruments potentially contaminated with the CJD agent are primarily derived from in vitro inactivation studies that used either brain tissues or tissue homogenates, both of which pose enormous challenges to any sterilization process.

The World Health Organization (WHO) has developed CJD infection control guidelines that can be a valuable guide to infection control personnel and other health care workers involved in the care of CJD patients. Destruction of heat-resistant surgical instruments that come in contact with high infectivity tissues, albeit the safest and most unambiguous method as described in the WHO guidelines, may not be practical or cost effective.

One of the three most stringent chemical and autoclave sterilization methods outlined in Annex III of the WHO guidelines (see below) should be used to reprocess heat-resistant instruments that come in contact with high infectivity tissues (brain, spinal cord, and eyes) and low infectivity tissues (cerebrospinal fluid, kidneys, liver, lungs, lymph nodes, spleen, olfactory epithelium, and placenta) of patients with suspected or confirmed CJD. High and low infectivity tissues were defined on the basis of available experimental data as described in Table 2 of the WHO guidelines. The stringent sterilization methods described below should be used to reprocess medical instruments that come in contact with high infectivity tissues of persons known to be blood relatives of patients with inheritable forms of TSEs. In addition, instruments should be kept moist and not allowed to air dry throughout the surgical procedure by immersing them in water or disinfectant solution.

What are the most stringent chemical and autoclave sterilization methods outlined in Annex III of the WHO infection control guidelines for transmissible spongiform encephalopathies?

The three most stringent sterilization methods for heat-resistant instruments described in Annex III of the WHO guidelines are listed below; the methods are listed in order of more to less severe treatments. Sodium hypochlorite may be corrosive to some instruments, such as gold-plated instruments. Before instruments are immersed in sodium hypochlorite, the instrument manufacturer should be consulted about the instrument’s tolerance of exposure to sodium hypochlorite. 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.

1. Immers 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 condensing on the lid of the container, 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. An experiment conducted by Food and Drug Administration (FDA) investigators indicated that the use of appropriate containment pans and lids prevents escape of sodium hydroxide vapors that may cause]
2. Immerse in 1N NaOH or sodium hypochlorite (20,000 ppm available chlorine) for 1 hour; clean; and subject to routine sterilization.

[ CDC NOTE: Sodium hypochlorite may be corrosive to some instruments. ]

3. Immerse 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.

[ CDC NOTE: Sodium hypochlorite may be corrosive to some instruments. ]

FDA investigators evaluated the effects to surgical instruments of the steps involved in the sterilization protocols listed above; some of the protocols they assessed subjected the instruments to harsher conditions than those prescribed above. Their findings indicate that much of the damage from autoclaving in sodium hydroxide was cosmetic and would not affect the performance or cleaning of the instruments. Soaking in sodium hydroxide had the least damaging effect on instruments, but immersion in sodium hypochlorite bleach caused severe damage to some instruments. The article summarizing the findings of this experiment by Brown et al. of the FDA was published in the Journal of Biomedical Materials Research (electronic version published October 2004) PDF version (191 KB/5 pages).

How should instruments used in patients with no clear diagnosis at the time of a neurosurgical procedure be reprocessed?

In some patients undergoing neurosurgery, a CJD diagnosis that is not suspected before the procedure may be confirmed after the neurosurgery. For this group of 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. Unless a clear non-CJD diagnosis is established, these patients should be considered as potentially suspected CJD patients for all other infection control requirements.

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

All disposable instruments, materials, and wastes that come in contact with high infectivity tissues (brain, spinal cord, and eyes) and low infectivity tissues (cerebrospinal fluid, kidneys, liver, lungs, lymph nodes, spleen, and placenta) of suspected or confirmed TSE patients should be disposed of by incineration. 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 with water.

[ CDC NOTE: Sodium hypochlorite may be corrosive to some instruments. ]

What kinds of precautions should be taken while embalming the bodies of patients with suspected or confirmed CJD?

An autopsied or traumatized body of a suspected or confirmed CJD patient can be embalmed, using the precautions outlined in the WHO CJD infection control guidelines. CJD patients who have not been autopsied or whose bodies have not been traumatized can be embalmed using Standard Precautions. Family members of CJD patients should be advised to avoid superficial contact (such as touching or kissing the patient's face) with the body of a CJD patient who has been autopsied. However, if the patient has not been autopsied, such contact need not be discouraged.
Additional recommendations for CJD infection control practices not addressed in this Q&A

Obtain them from: