Frequently Asked Questions about Creutzfeldt-Jakob Disease (CJD)

Prepared by the North Carolina Division of Public Health

GENERAL INFORMATION

What is Creutzfeldt-Jakob disease (CJD)?

CJD is a rapidly progressive, fatal brain disease related to abnormal proteins called prions. The majority of CJD patients die within one year of illness onset. CJD is rare and occurs with an estimated annual incidence of about one case per million persons worldwide. About 200–300 confirmed cases of CJD are reported each year in the United States.[1]

What are the types of CJD?

- **Sporadic CJD:** In about 85 percent of patients, CJD occurs sporadically with no recognizable source of infection. This most common form of CJD typically affects adults between 55 and 75 years old.
- **Familial or genetic CJD:** About 10-15 percent of CJD cases are associated with inherited mutations in the prion gene.
- **Acquired CJD (includes both iatrogenic and variant CJD).**
    - **Iatrogenic CJD:** Iatrogenic transmission (i.e. transmission through medical treatment) is rare, accounting for less than one percent of CJD cases. Most iatrogenic CJD cases have been linked to use of hormones or tissues obtained from persons with undiagnosed CJD. Six cases of CJD from the late 1950s to 1976 were linked to prion-contaminated neurosurgical instruments and devices; four associated with contaminated neurosurgical instruments and two with implantable electroencephalogram depth electrodes.[2]
    - **Variant CJD:** Variant CJD (vCJD) was first identified in 1996 in the United Kingdom in persons who had eaten contaminated meat from cattle infected with Bovine Spongiform Encephalopathy (BSE), also known as Mad Cow Disease. Although three cases of vCJD have been reported in the United States, all were in persons who were exposed to BSE while living in other countries.

Have CJD cases been reported in North Carolina?

Yes. Reporting of CJD has been required in North Carolina since August of 2002. Healthcare providers are required to report suspected or confirmed cases of CJD to the local health department within seven days after the disease is reasonably suspected to exist. From 2008 through 2013, there have been 51 confirmed cases of CJD reported in North Carolina (average 8.5 cases per year, or approximately one case per million residents). During this same period, there have also been 37 probable cases of CJD reported, i.e., cases that could not be confirmed but had clinical findings consistent with CJD.

What is the incubation period for acquired CJD?

The incubation period (time from exposure to symptom onset) for acquired CJD is not well known, but appears to be very long (i.e., years to decades).

What are the symptoms of CJD?

In the early stages of CJD, the patient may have rapidly failing memory and other cognitive difficulties. Other symptoms can include personality changes such as anxiety and depression; lack of coordination; and visual disturbances. As the illness progresses, mental deterioration becomes pronounced and other symptoms can appear, including sudden jerky movements; blindness; weakness of the limbs; and coma. Once symptoms begin, the average time to death is four months but may be as long as 12 months.

How is CJD diagnosed?

Several tests can be used to help diagnose CJD. Cerebrospinal fluid (CSF) may be taken for testing and other studies such as electroencephalogram (EEG) and magnetic resonance imaging (MRI) may also be conducted. However, the only way to confirm the diagnosis of CJD is by examining brain tissue. No tests are available to determine the risk in a person potentially exposed through use of neurosurgical instruments that had been used for a person with CJD.

Is there a cure for CJD?
No. CJD is always fatal.

Can CJD be treated?
No specific treatments are available for CJD. However, measures can be taken to reduce the patient's symptoms and provide comfort.

How is CJD transmitted?
The vast majority of CJD cases are sporadic or familial; less than one percent of all CJD cases are transmitted from another source. CJD can be transmitted by exposure to brain tissue, spinal cord fluid, and certain other tissues from an infected person. Historically, the following circumstances have been associated with transmission of CJD:

- Injection with cadaver-derived pituitary hormones, dura mater and corneal grafts from cadavers of patients infected with CJD, and
- Exposure to CJD-contaminated instruments during neurosurgical or ophthalmic surgical procedures.

CJD cannot be transmitted through the air by sneezing or coughing. CJD cannot be transmitted by casual contact like touching, kissing, or even by sexual contact.

Have there been any incidents of potential exposure to CJD through neurosurgical procedures in the United States?
Yes. According to the Centers for Disease Control (CDC), there were 19 incidents of patient exposures to surgical instruments potentially contaminated with CJD during 1998–2012, including 17 that involved neurosurgical procedures and two that involved ophthalmologic procedures.[2] At least two other potential exposures have been reported since 2012. To date, none of the patients exposed in these incidents have developed CJD.

Have there been any incidents of potential exposure to CJD through neurosurgical procedures in North Carolina?
Yes. On February 10, 2014, Novant Health Forsyth Medical Center announced that 18 patients had been exposed to neurosurgical instruments used on a patient who was later confirmed to have CJD. The instruments used during the patient's surgery were sterilized using standard procedures for cleaning surgical instruments, but they were not subjected to enhanced sterilization procedures recommended for CJD.

What are the chances of acquiring CJD during surgery?
The risk of developing any type CJD is very small, since only one case is reported per million persons each year. Of all CJD cases reported worldwide, less than one percent are known to be transmitted through medical procedures. Transmission from exposure to surgical instruments contaminated with CJD has never been documented in the United States.

INFORMATION FOR HEALTHCARE PROVIDERS

How is the diagnosis of CJD confirmed?
The only way to confirm the diagnosis of CJD is by examining brain tissue. Sometimes the diagnosis may be confirmed through brain biopsy, but the affected area may be missed. **The National Prion Disease Pathology Surveillance Center** conducts testing for CJD; Autopsy Coordinators are available at 216-368-0587 for consultation.

Where can healthcare providers receive diagnostic support?
The National Prion Disease Pathology Surveillance Center is available for physician diagnostic consultation at 216-368-0587. The National Prion Disease Pathology Surveillance Center is located at Case Western University and is supported by the CDC and sponsored by the American Association of Neuropathologists.

Are healthcare providers required to report CJD to public health?
Yes. CJD is a reportable condition under the North Carolina Administrative Code rules. Providers who suspect CJD should contact their local health department within seven days after the diagnosis of CJD is reasonably suspected. Reporting should not be delayed while awaiting laboratory or pathology findings.

Why is infection prevention awareness important and different for CJD?
Prions are resistant to conventional sterilization methods. To prevent CJD exposures, CDC recommends that CJD infection prevention guidelines be developed and implemented by each healthcare facility that performs neurosurgical or ophthalmologic surgical procedures.

How do hospitals prevent transmission of CJD?
Guidance for infection control personnel and other health care workers involved in the care of CJD patients is available from the World Health Organization (WHO) and from the Society for Healthcare Epidemiology of America.[3,4] Fundamental tenets of CJD infection prevention include the following:

- The possibility of CJD should be considered as part of the patient's pre-surgical assessment prior to an intracranial procedure.
- Neurosurgical instruments used in procedures on patients with an unclear diagnosis (particularly instruments used for brain biopsies) should be regarded as potentially contaminated with CJD. Such instruments should be quarantined until a nonprion disease diagnosis is identified or should be regarded as contaminated and sterilized using recommended CJD decontamination protocols.[3-5]

How should instruments used in patients with no clear diagnosis at the time of a neurosurgical procedure be reprocessed?
Several potential exposure incidents have occurred when the diagnosis of CJD was not confirmed or even considered until after the patient had already undergone neurosurgery.[2] For this reason, neurosurgical instruments used for patients with unclear diagnoses should be regarded as potentially contaminated with CJD and should either be quarantined until a nonprion disease diagnosis is identified, or sterilized using recommended CJD decontamination protocols.[3-5]

How should heat-sensitive instruments or materials or surfaces 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 CJD 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 (20,000 ppm) for one hour and rinsed with water.[5] (NOTE: Sodium hypochlorite may be corrosive to some instruments.)

Should hospitals track the use of surgical equipment to determine which instruments were used for which patients?
Yes. The feasibility of implementing instrument tracking procedures should be considered, since this could reduce confusion in the event that a notification is required.

CJD RESOURCES

Centers for Disease Control and Prevention:

- Main CJD website: http://www.cdc.gov/ncidod/dvrd/cjd/index.htm
- Infection Control Practices: http://www.cdc.gov/ncidod/dvrd/cjd/qa_cjd_infection_control.htm
- Information for Funeral Directors: http://www.cdc.gov/ncidod/dvrd/cjd/funeral_directors.htm

National Prion Disease Pathology Surveillance Center: http://www.cjdsurveillance.com/protocols.html

North Carolina Administrative Code Rule 10A NCAC 41A .0212: Handling and Transportation of Bodies.

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