

The 2019 Centers for Disease Control and Prevention Report

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CDC Fast Facts



- ❑ CDC operates under the Department of Health and Human Services (DHHS) and is the nation's leading public health agency.
- ❑ Attempts to keep America secure by controlling disease outbreaks, making sure food and water are safe, helping people to avoid leading causes of death, and working globally to reduce threats to the nation's health.
- ❑ Headquartered in Atlanta with facilities in 10 additional U.S. locations; field staff in all states and more than 50 countries.
- ❑ More than 14,000 employees in nearly 170 occupations

Office and Mission

- National Center for Emerging and Zoonotic Infectious Diseases
 - Division of High -Consequence Pathogens and Pathology
 - Prion and Public Health Office
- Surveillance (for human prion disease)
 - Monitoring of disease in population (e.g., estimation of prion disease rates, detection of changes in epidemiology of disease over time, monitoring of possible occurrence of variant CJD (vCJD) or novel prion diseases, gaining of knowledge about prion diseases)

Surveillance: Prion Diseases

- ❑ To estimate prion disease incidence in the US, we match death certificate data with data from the National Prion Disease Pathology Surveillance Center (NPDPSC).
 - National multiple cause-of-death data (death certificate data) is compiled by CDC's National Center for Health Statistics (NCHS).
 - Routinely obtained and costeffective
 - Good source of information because of disease fatality rate (100%); diagnosis more accurate at late stages of disease
 - If CJD, prion disease, GSS, etc. is listed anywhere on the death certificate, it is included in the NCHS data (misspellings, too).
 - Results of specimen testing by NPDPSC may confirm or rule out suspected prion disease cases
 - Cases are added to or subtracted from death certificate data based on NPDPSC information

Surveillance: Prion Diseases

- ❑ **5737 decedents were identified as having prion disease during 2003-2016 for an average annual age-adjusted incidence of 1.2 cases per million population.**
 - The incidence among males was 1.3 per million, and among females, 1.1 per million.
 - Incidence among white decedents (1.3 per million) was more than double the incidence among black decedents (0.6 per million).
- ❑ **12 cases were <30 years of age: 6.9 per billion per year.**
 - Only 2 were sporadic forms of prion disease, 1 sporadic CJD and 1 sporadic fatal insomnia, for an annual incidence of 1.1 per billion.
 - The remaining 10 cases were familial (4 GSS, 2 FFCJD), variant CJD (2), and iatrogenic CJD (1, dura mater associated).
- ❑ **Incidence among those ≥ 65 : 5.9 per million per year.**
- ❑ **1 CJD death for approximately every 6,000 deaths overall in the US each year**

Surveillance: Prion Diseases

- Approximately 12% (range 7% -18%) of the total cases each year during 2003 -2016 were the result of additions to the death certificate data based on positive neuropathologic and/or genetic findings.
- Approximately 5% (range 3% -6%) of the death certificate cases were removed annually based on negative neuropathology results.
 - NPDPSA's now regular use of the highly specific (few false positives), pre mortem RT-QuIC test *should* lead to improvements in death certificate accuracy in the coming years.
 - CDC case definition: Neuropsychiatric illness + positive RT-QuIC = probable CJD

Surveillance: Disease Confirmation

- ❑ Prion disease diagnoses *still* confirmed only by neuropathology
 - Neuropathology can be a sentinel for unique and possibly emerging prion disease cases.
- ❑ RT-QuIC continues to be assessed through comparison of results with the “gold standard” of brain tissue analysis.
- ❑ NPDPSC neuropathology results are used to adjust death certificate data, allowing for more accurate incidence calculations.

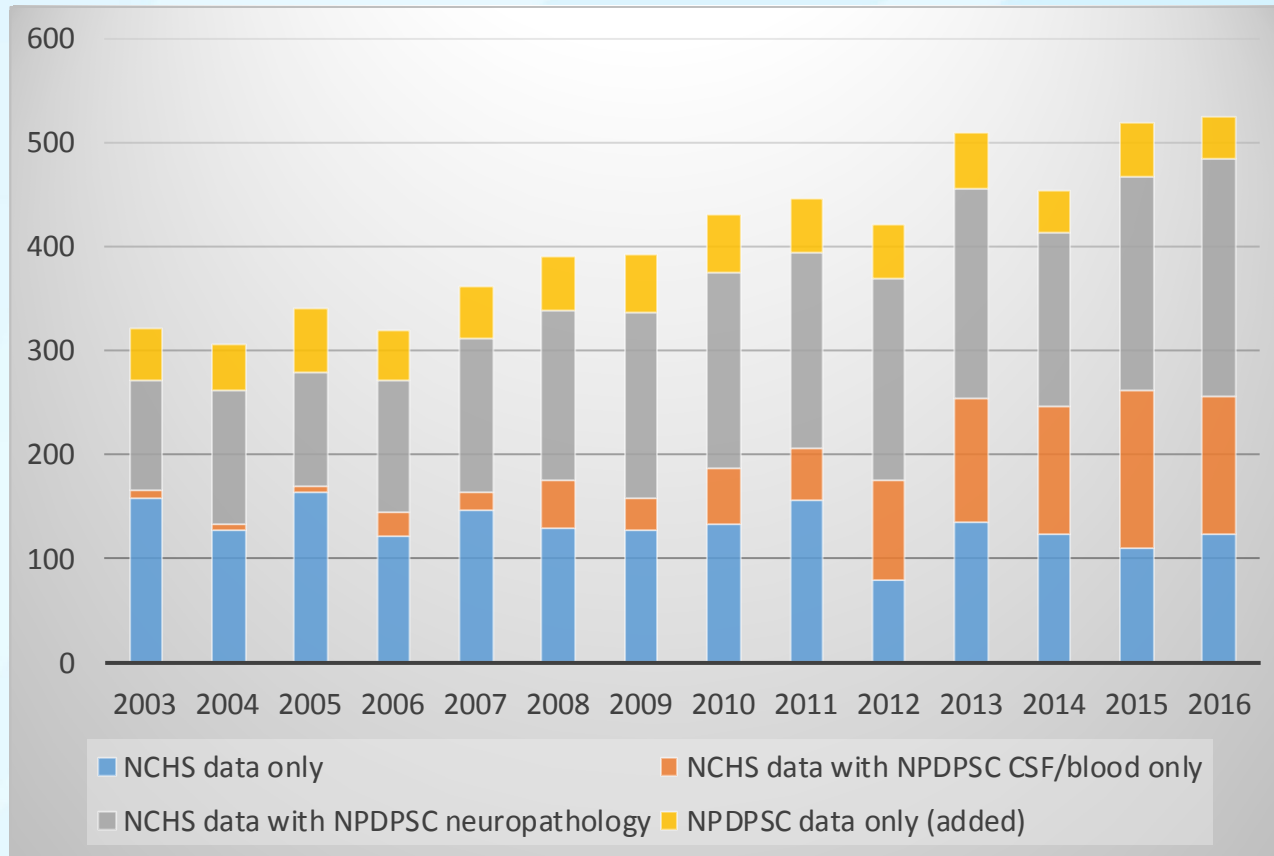
Surveillance: Mechanisms

- ❑ CDC may learn of a CJD case through a variety of different sources:
 - NCHS (national multiple cause-of-death data)
 - National Prion Disease Pathology Surveillance Center (NPDPSC)
 - Public health departments and medical personnel
 - Family members, the public, and the media
- ❑ Cause of death on death certificates *can* be amended; the process varies from state to state.
- ❑ Verified cases are included as part of our national prion disease surveillance.
- ❑ Surveillance does *not* capture every CJD case.

Surveillance: States

- ❑ CJD is now reportable to some degree in almost all states.
- ❑ State reporting requirements do not necessarily translate into more accurate surveillance.
 - Misdiagnosed case will still be a misdiagnosed case.
- ❑ CDC helped co-author a Council of State and Territorial Epidemiologists (CSTE) position statement outlining specific CJD surveillance actions and goals.
 - Making the disease reportable in a state may facilitate accomplishment of these goals.
- ❑ CDC provides funding to strategic states for enhanced surveillance activities.

Prion disease cases by year, United States, 2003-2016



Surveillance: Variant CJD



- ❑ Variant CJD is the human form of bovine spongiform encephalopathy (BSE, or “mad cow disease”).
- ❑ 231 cases of variant CJD worldwide (178 in United Kingdom)
- ❑ 4 cases in the United States, 2 in Canada
 - None are believed to have been exposed to the infectious agent in North America.

Organ, Tissue, and Blood Donation

- FDA Guidance for Industry: Blood, human cells, tissue, and cellular and tissue -based products (corneas, skin, bone, heart valves, etc.)

Ineligible:

- Persons who have been diagnosed with vCJD or any other form of CJD
- Persons who have a history of CJD in a blood relative
- Persons who spent three months or more cumulatively in the United Kingdom from the beginning of 1980 through the end of 1996
- Persons who spent 5 years or more cumulatively in Europe from 1980 until the present

Organ, Tissue, and Blood Donation (continued)

- ❑ **CJD has not been shown to be transmissible through blood.**
 - American Red Cross and CDC (2017): “From this study, as well as other epidemiologic studies, there is no evidence of CJD transfusion transmission; this risk remains theoretical.”
 - United Kingdom National CJD Research and Surveillance Unit (2017): “The balance of evidence indicates that sCJD is transmitted by blood transfusion, it must be a rare event, if it happens at all...”
- ❑ **Variant CJD *has* been transmitted through blood in a few cases.**

Organ, Tissue, and Blood Donation (continued)



- ❑ FDA does *not* regulate organ donation (liver, kidney, etc.)
 - There are no absolute exclusions.
- ❑ In other words, organs from those with CJD *can* be donated, BUT organs may or may not be used depending on outcome of risk-benefit analysis by medical staff.

<http://www.fda.gov/downloads/biologicsbloodvaccines/guidancecomplianceregulatoryinformation/guidances/tissue/ucm091345.pdf>

<http://www.fda.gov/downloads/BiologicsBloodVaccines/GuidanceComplianceRegulatoryInformation/Guidances/Blood/UCM307137.pdf>

Funeral Homes

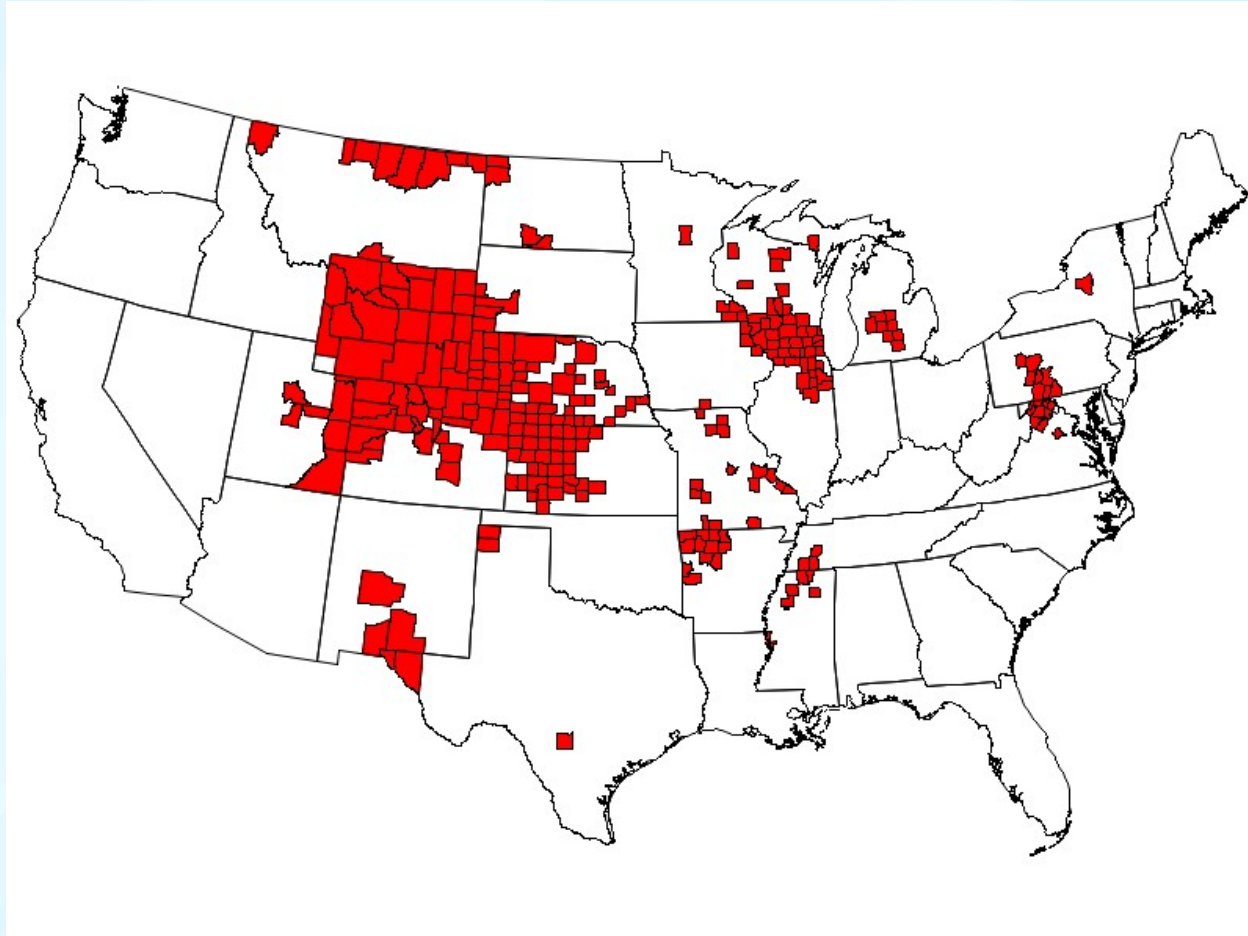
- ❑ Embalming bodies of CJD patients who have been autopsied can be safely performed, BUT a funeral home cannot be forced to accept a body.
- ❑ Education of funeral directors is important; however, some will be more open to revising policies than others.
- ❑ Information for funeral and crematory practitioners is available on the CDC website.
- ❑ “There are no special interment, entombment, inurnment, or cremation requirements for patients with CJD. Interment of bodies in closed caskets does not present a significant risk of environmental contamination and cremated remains can be considered sterile, as the infectious agent does not survive incineration -range temperatures.”

Chronic Wasting Disease (CWD)



- ❑ Prion disease of cervids, including white -tailed deer, mule deer, elk, moose, and reindeer
- ❑ Can be highly transmissible within cervid populations
- ❑ Found among free -ranging deer and elk in 24 states (22 identified within past two decades) and 2 Canadian provinces
 - Recently reported in Norway (2016), Finland (2018), and Sweden (2019).

Chronic Wasting Disease Among Free-Ranging Cervids by County, United States, June 2019



Chronic Wasting Disease

'Zombie' deer disease is in 24 states and thousands of infected deer are eaten each year, expert warns

Ryan W. Miller, USA TODAY Published 10:15 a.m. ET Feb. 16, 2019 | Updated 2:13 p.m. ET Feb. 18, 2019



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ANIMALS

The Dangers Of 'Zombie Deer Disease'

May 11, 2019 · 8:16 AM ET
Heard on Weekend Edition Saturday

- ❑ To date, there is no strong epidemiologic evidence for the occurrence of CWD in people.

CWD Studies

- ❑ **Goal: To determine whether CWD can cause disease in humans**
 - ❑ **Follow-up of persons who hunted in Wyoming and Colorado, where CWD has been present for years, and identifying those who died of prion disease**
 - ❑ **Follow-up of individuals who consumed venison from CWD - positive deer in Wisconsin**
 - **Prion 2019 poster presentation: To date, no matches found among potentially exposed persons who were crosschecked with NPDPSC and national multiple cause-of-death data**
 - ❑ **Analysis of highly CWD-endemic states compared to non -CWD states to assess whether differences in the rate of human prion disease mortality exist.**

Final Thoughts

- ❑ Prion diseases present a unique diagnostic and public health challenge.
- ❑ CDC's prion disease-related activities include:
 - conducting surveillance through various methods to best capture the majority of cases.
 - investigating cases of interest in collaboration with affected states.
 - providing advice on prion disease-related issues.
- ❑ Collaboration with medical and public health personnel, NPDPS, and the CJD Foundation is essential.
- ❑ Improvements in pre-mortem diagnostic testing (i.e., RT-QuIC) should benefit surveillance efforts; however, autopsy remains important.

Resources

❑ CJD Foundation

- 1-800-659-1991
- www.cjdfoundation.org

❑ Centers for Disease Control and Prevention

- 404-639-3091
- <http://www.cdc.gov/prions/cjd/index.html>

❑ National Prion Disease Pathology Surveillance Center

- 216-368-0587
- <http://case.edu/med/pathology/centers/npdpssc/>

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 - Ms. Marissa Person
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 - CJD Foundation
 - NPDPSC
 - State and local public health departments

Questions?

For more information please contact Centers for Disease Control and Prevention

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The findings and conclusions in this report are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention.

