

# The Centers for Disease Control and Prevention Report: A CDC CJD Q&A

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Division of High-Consequence Pathogens and Pathology



# What is CDC, and how is it involved with prion diseases?



## **CDC Fast Facts**

- ❑ **CDC is one of the major operating components of 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 such as heart disease and cancer; and working globally to reduce threats to the nation's health.**
- ❑ **Headquartered in Atlanta with facilities in 10 additional U.S. locations**
- ❑ **More than 14,000 employees in nearly 170 occupations**
- ❑ **Field staff works in all 50 states and more than 50 countries**

# CDC's Responsibility

- **The DHHS BSE/TSE Action Plan has four major components:**
  - **Oversight:** primarily the responsibility of the Office of the Secretary of DHHS
  - **Protection:** primarily the responsibility of the Food and Drug Administration (FDA)
  - **Research:** primarily the responsibility of the National Institutes of Health (NIH)
  - **Surveillance (for human disease):** primarily the responsibility of CDC
    - 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)

**How rare is this disease *really*?**



## **Death certificate data**

- ❑ **The National Center for Health Statistics (NCHS) compiles national multiple cause-of-death data.**
- ❑ **Death certificate data review is effective as a surveillance tool for CJD:**
  - 100% fatality rate
  - Diagnosis more accurate at late stages of disease
  - Active review has shown high ascertainment rate
  - Mortality data are routinely obtained by NCHS on an ongoing basis; use for CJD surveillance cost-effective

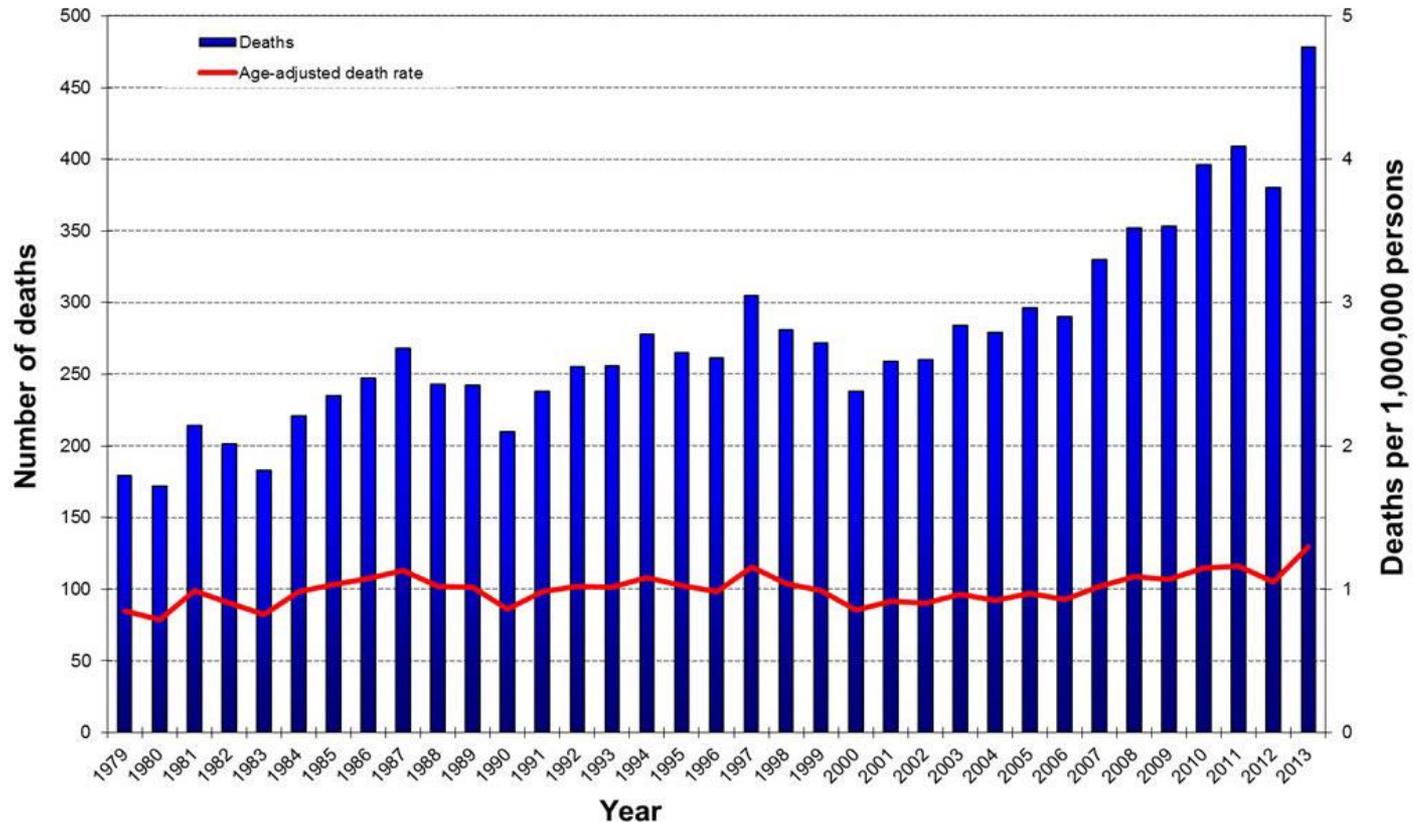
## One in a million?

- ❑ **CJD occurs worldwide at a rate of about 1 – 1.5 per million population per year, although rates of up to 2 cases per million are not unusual.**
- ❑ **To more accurately determine incidence in the United States, we attempt to match death certificate data with National Prion Disease Pathology Surveillance Center (NPDPSC) data.**
  - Based on NPDPSC neuropathology results, cases are added to or subtracted from death certificate data.
- ❑ **For 2003-2011, almost half (49.0%) of the death certificates matched with a diagnosed decedent in the NPDPSC database with neuropathological or genetic test results.**
- ❑ **For this time period, the matching process yielded an incidence rate of 1.15 cases per million.**

## But...

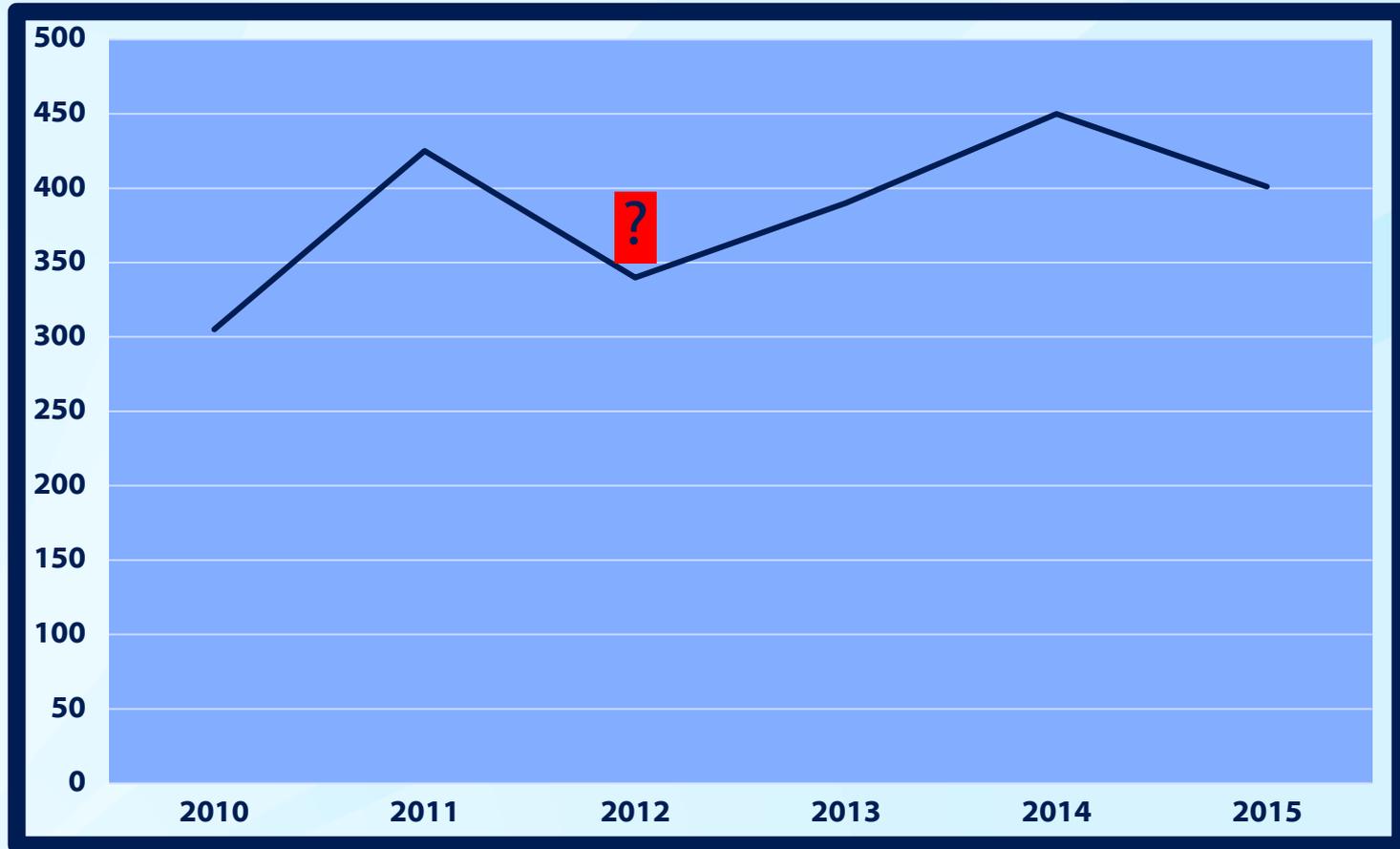
- ❑ The average annual CJD incidence rate among decedents  $\geq 65$  years of age (2008-2010): 6.3 cases per million.
- ❑ Incidence of sporadic prion disease among decedents  $< 30$  years of age (1979-2014): 1.6 per *billion*.
- ❑ 1 CJD death for approximately every 6,000 deaths overall in the US each year

# Creutzfeldt-Jakob disease deaths and age-adjusted death rate, United States, 1979-2013\*



\* Deaths obtained from the multiple cause-of-death data for 1979-1998 are based on ICD-9 codes, and those beginning in 1999 are based on ICD-10 codes with available computerized literal death certificate data. Death information was also obtained from other surveillance mechanisms; data include familial prion diseases. Rates are adjusted to the US standard 2000 projected population.

## Was my loved one's death *counted*?



## 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
- ❑ **Verified cases are included as part of our national prion disease surveillance.**
- ❑ **Surveillance does *not* capture every CJD case.**

## Death certificates

- ❑ **If CJD, prion disease, GSS, etc. is listed anywhere on the death certificate, it is included in the NCHS data.**
  - Misspellings, too!
- ❑ **Cause of death on death certificates *can* be amended.**
  - The process varies from state to state.
  - Some states require that the certifying physician or medical examiner make this amendment.
- ❑ **If there is no indication of prion disease anywhere on the death certificate, we still may be aware of the case through NPDPS data or other sources mentioned previously.**

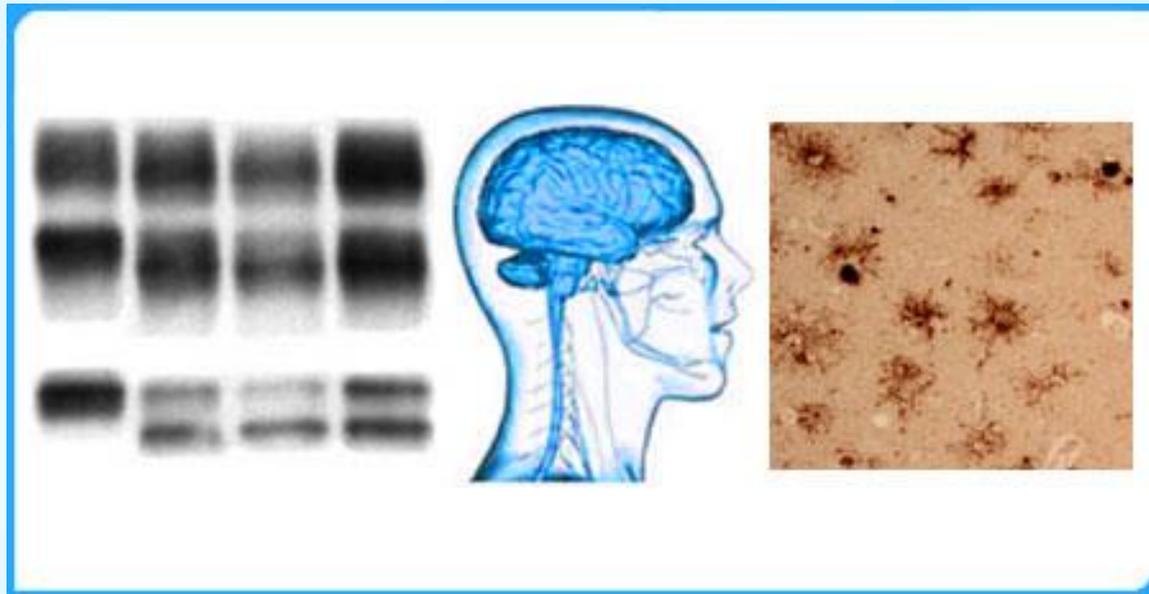
# Is CJD reportable in my state? Should it be?



## State reporting

- ❑ **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.

# Is an autopsy still necessary? Can my loved one be a donor?



# Autopsy

- ❑ **Prion disease diagnoses *still* confirmed only by neuropathology**
  - Neuropathology can be a sentinel for unique and possibly emerging prion disease cases.
- ❑ **New and improved premortem diagnostic tests such as RT-QuIC can be validated 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.**

# 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 (unless no mutation is present)
  - 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
- ❑ **CJD has not been shown to be transmissible through blood: however, vCJD *has* been transmitted through blood in a few cases.**
  - ❑ **It is recommended that organs from those with known or suspected CJD should not be donated.**

<http://www.fda.gov/downloads/biologicsbloodvaccines/guidancecomplianceregulatoryinformation/guidances/tissue/ucm091345.pdf>  
<http://www.fda.gov/downloads/BiologicsBloodVaccines/GuidanceComplianceRegulatoryInformation/Guidances/Blood/UCM307137.pdf>

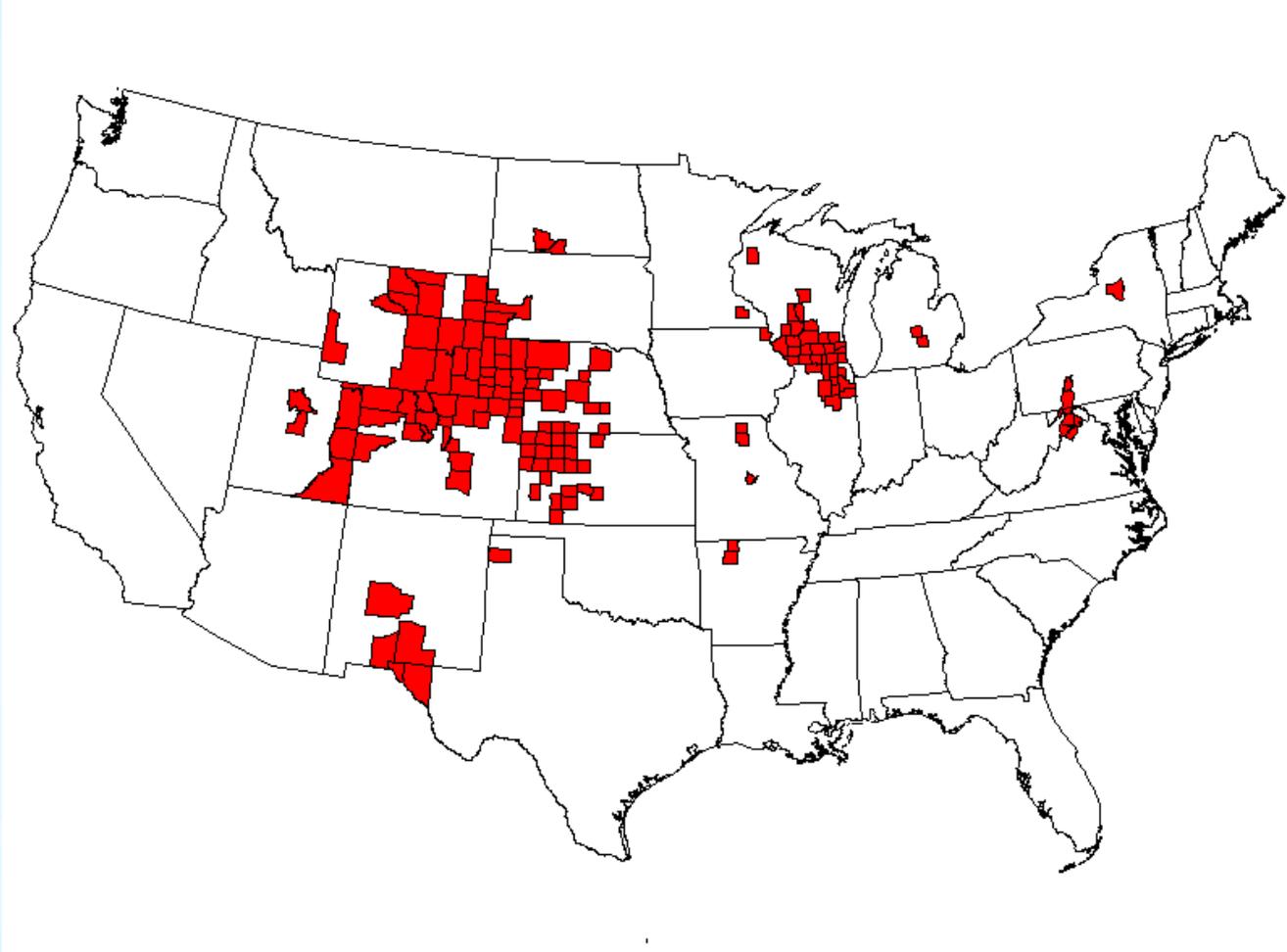
## Funeral homes

- ❑ **Embalming bodies of CJD patients who have been autopsied can be safely performed. Unfortunately, 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.”**

# Should I be concerned about getting chronic wasting disease (CWD)?



# Chronic Wasting Disease Among Free-Ranging Cervids by County, United States, April 2016



## **Hunter studies**

- ❑ **Goal: To determine whether CWD, a prion disease of deer and elk (and moose, and reindeer...), can cause disease in humans**
- ❑ **Follow-up of persons who hunted in Wyoming and Colorado, where CWD is found, and identifying those who died of prion disease**
  - Results: Prion disease cases among these groups within expected range so far, but many years of follow-up necessary
- ❑ **Follow-up of hunters who consumed venison from CWD-positive deer in Wisconsin**

**How many variant CJD cases have there been in the United States? How do you know they weren't exposed here?**

## **Recent US Case of Variant Creutzfeldt-Jakob Disease—Global Implications**

**Atul Maheshwari, Michael Fischer, Pierluigi Gambetti, Alicia Parker, Aarthi Ram, Claudio Soto, Luis Concha-Marambio, Yvonne Cohen, Ermias D. Belay, Ryan A. Maddox, Simon Mead, Clay Goodman, Joseph S. Kass, Lawrence B. Schonberger, Haitham M. Hussein**



## **Variant CJD**

- ❑ **Variant CJD is the human form of bovine spongiform encephalopathy (BSE, or “mad cow disease”).**
- ❑ **Four cases in the United States, and two in Canada, have been identified.**
  - None are believed to have been exposed to the infectious agent in North America based on epidemiological evidence including the decedents’ countries of origin, travel histories, and current knowledge about incubation periods and susceptibility factors associated with the disease.

## **Final thoughts**

- ❑ **CJD presents a unique diagnostic and public health challenge.**
- ❑ **CDC conducts surveillance for prion diseases through various methods to best capture the majority of cases.**
- ❑ **CDC investigates cases of interest in collaboration with affected states.**
- ❑ **CDC provides advice on prion disease-related issues.**

## **Final thoughts**

- ❑ Collaboration with medical and public health personnel, NPDPSC, the CJD Foundation, and CDC is essential.**
- ❑ Future surveillance will be helped by increased autopsy rates, improved pre-mortem diagnostic tests, and physician awareness of NPDPSC's services.**

# CJD resources

## ❑ CJD Foundation

- 1-800-659-1991
- [www.cjdfoundation.org](http://www.cjdfoundation.org)

## ❑ Centers for Disease Control and Prevention: Division of High-Consequence Pathogens and Pathology

- 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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## □ All the wonderful people at:

- CJD Foundation
- NPDPS
- State and local public health departments

# Other 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.

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