# The 2023 Centers for Disease Control and Prevention (CDC) Report

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CJD Foundation Family Conference

# CDC Fast Facts

- CDC operates under the Department of Health and Human Services and is the nation's leading public health agency, working to:
  - **Control disease outbreaks**
  - **Make sure food and water are safe**
  - **Help people avoid leading causes of death**
  - **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

# CDC Organization

- National Center for Emerging and Zoonotic Infectious Diseases
  - Division of High-Consequence Pathogens and Pathology
    - Prion and Public Health Office (PPHO)

# PPHO Mission

- **Surveillance (for <u>human</u> 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 cost-effective
    - 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

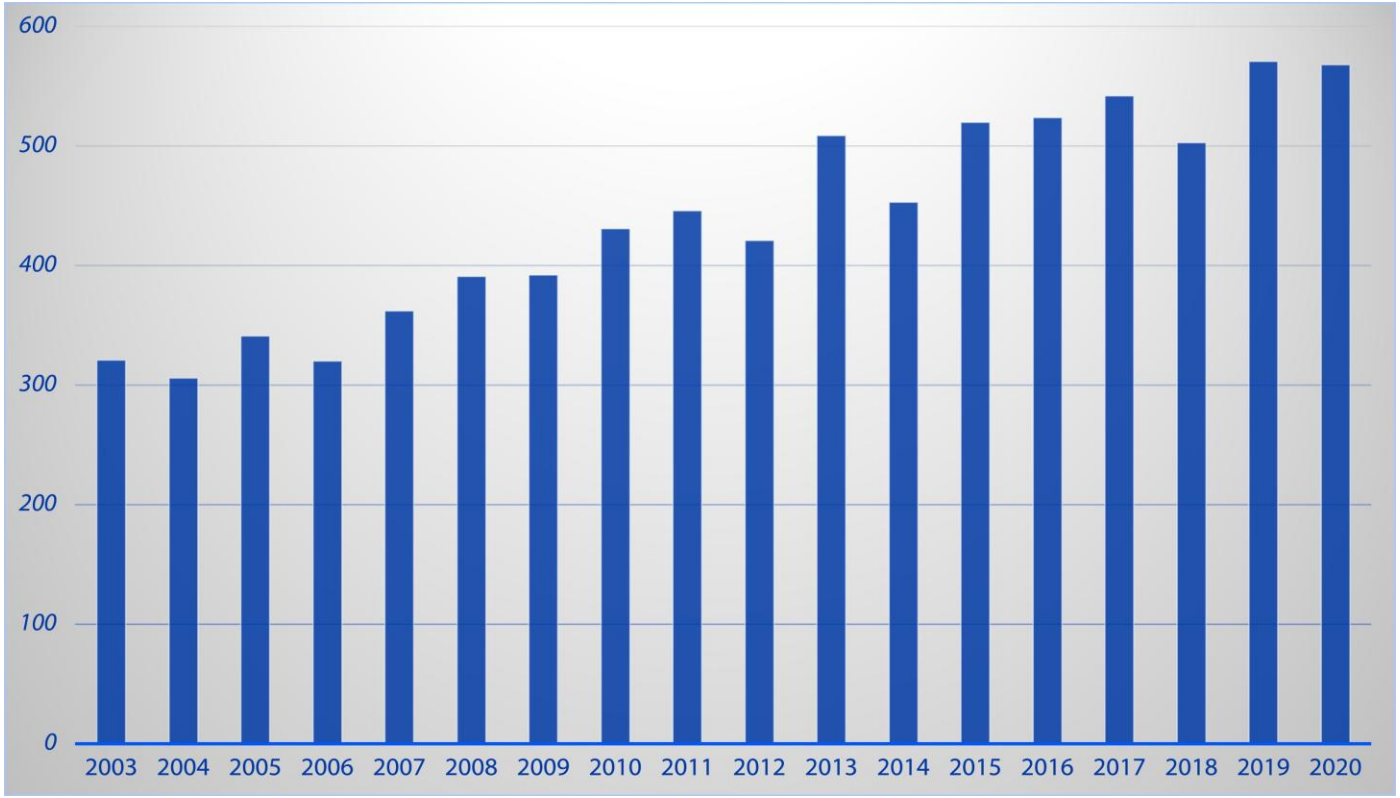
# Surveillance: Prion Diseases

- 7921 decedents were identified as having prion disease during 2003-2020 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 those ≥65: 6.1 cases per million per year.
- 1 CJD death for approximately every 6,000 deaths overall in the US in 2020

# Surveillance: Prion Diseases

- 2020 data:  568 cases (compared to 571 cases in 2019); average age-adjusted incidence: 1.3 per million population
- Definite or highly probable matches for 87% of cases in the national death certificate data when compared to diagnostic testing data from NPDPSC
  - **This high percentage is a testament to NPDPSC's expertise and the increased awareness of its services.**
- 10 cases in the death certificate data were found to have negative NPDPSC autopsy results and were removed from the total; 40 autopsy-confirmed cases were identified in the NPDPSC data with a death in 2020 that did not match with a case in the death certificate data and were added.

# Prion Disease Cases by Year, United States, 2003-2020



# Surveillance: RT-QuIC

- An RT-QuIC-positive test result strongly indicates prion disease.
- If NPDPSC does not conduct autopsy analyses on an RT-QuIC-positive case, date of death data for the patient may not be available.
- A number of such cases were identified in the NPDPSC database; we estimate they could account for ~20% more cases in 2020 based on the dates that NPDPSC received the samples for testing.
- Proposed next step: Using the National Death Index to identify dates of death for RT-QuIC-positive cases in the NPDPSC data and then adding these cases to the appropriate years to obtain an even more accurate estimate of national prion disease incidence.

# Surveillance: Young Cases (<30 years)

- Most young cases have a genetic mutation or other risk factor (human growth hormone, vCJD)
- **18 young cases 2003-2020: 8 cases per *billion* per year.**
  - Only 6 were sporadic forms of prion disease: 3 sporadic CJD and 3 sporadic fatal insomnia.
  - The remaining 12 cases were familial (5 GSS, 2 FFI, 2 fCJD), variant CJD (2), and iatrogenic CJD (1, dura mater-associated).
- Usually ≤ 1 case each year
  - 3 cases in 2019, 1 case in 2020
  - Preliminary findings: 1 case in 2021 (sFI), 0 in 2022

# 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.
  - **However, as mentioned earlier, a positive RT-QuIC in the context of a neuropsychiatric illness is <u>strongly</u> indicative of prion disease.**

# 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.

# Surveillance: Clusters

- Possible clusters of prion disease are occasionally reported to CDC.
- Fortunately, investigations inevitably “dissolve” the reported clusters, due to:
  - **Case(s) misdiagnosed or misreported as prion disease**
  - **Case(s) residing outside of the cluster area**
  - **Case(s) having a genetic form of prion disease**
  - **Case number actually within expected range given the population/region served by a hospital/number of years included, etc.**
- State and local health departments are valuable partners in these investigations

# Surveillance: Variant CJD (vCJD)

- Variant CJD is the human form of bovine spongiform encephalopathy (BSE, or “mad cow disease”).
- 233 cases worldwide (178 in U.K.)
- **4 cases in the United States, 2 in Canada (none believed to have been exposed to the infectious agent in North America).**
- **3 of the 4 vCJD cases reported since 2016 have been attributed to occupational exposure rather than consumption of contaminated beef.**

# Tissue and Organ Donation

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

  Ineligible:

  - **Persons diagnosed with vCJD or any other form of CJD**
  - **Persons who have a history of CJD in a blood relative**
  - **Persons who spent ≥ 3 months cumulatively in the United Kingdom from 1980-1996; persons who spent ≥5 years cumulatively in Europe 1980-present**
- FDA does *not* regulate organ donation (liver, kidney, etc.)
  - **There are <u>no</u> absolute exclusions (i.e., 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).**

# Blood Donation: New(ish) Recommendations (5/2022)

- Updated guidance:
  - **Removes donor deferral for geographic risk of BSE exposure**
- What does that mean?
  - **Donors previously deferred for time spent in the U.K., France, and Ireland, or for receipt of a blood transfusion in the U.K., France, or Ireland, may now be eligible if they meet all other eligibility requirements.**

https://www.fda.gov/media/124156/download

# Blood Donation: New(ish) Recommendations (5/2022)

- Still ineligible:
  - **Persons who have been diagnosed with vCJD, CJD, or any other transmissible spongiform encephalopathy or who have a blood relative diagnosed with familial prion disease (e.g., fCJD, GSS, or FFI)**
  - **Persons who received cadaveric pituitary hGH treatment**
  - **Persons who received a human cadaveric (allogeneic) dura mater transplant.**

# Blood Donation: Final Notes

- 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…”
  - U.K. National CJD Research and Surveillance Unit (2017): “The balance of evidence indicates that, if sCJD is transmitted by blood transfusion, it must be a rare event, if it happens at all…”
- **Variant CJD *has* been transmitted through blood.**
  - Reported in U.K. recipients of blood collected up to 3 years before vCJD onset in the donors.
- Because many U.K. residents have potentially been exposed to BSE, concerns still exist about additional secondary spread of the agent via blood and possibly contaminated surgical instruments.

# Funeral Homes

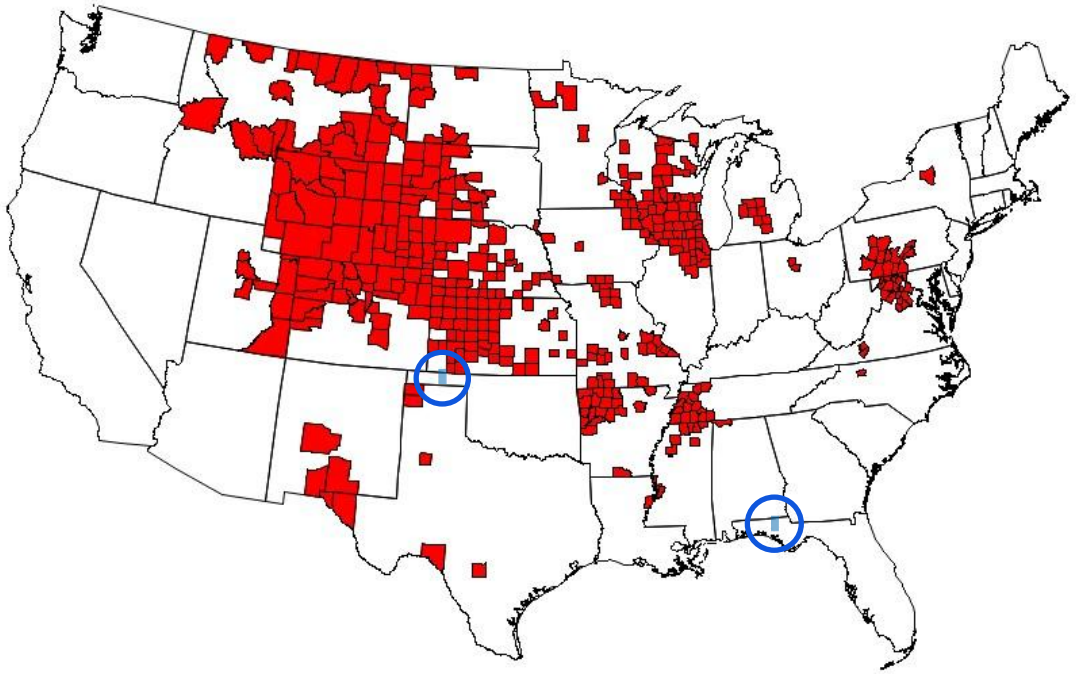
- Embalming bodies of CJD patients who have been autopsied can be safely performed, BUT a funeral home cannot be forced to accept a body.
- Education of funeral directors is important; however, some will be more open to revising policies than others.
- Information for funeral and crematory practitioners 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.”

http://www.cdc.gov/prions/cjd/funeral-directors.html

# Chronic Wasting Disease (CWD)

- Prion disease of cervids, including white-tailed deer, mule deer, elk, moose, and reindeer
- Clinical symptoms include weight loss, behavioral changes, excessive salivation, difficulty swallowing
- Can be highly transmissible within cervid populations
- **Found among free-ranging deer and elk in 31 states (Oklahoma and Florida this year) and 3 Canadian provinces**
- Also reported among free-ranging moose and/or reindeer in Norway (2016), Finland (2018), and Sweden (2019)

# Chronic Wasting Disease Among Free-Ranging Cervids by County, United States, July 2023



# CWD Transmission to Humans

- To date, there is no strong epidemiologic evidence for the occurrence of CWD in people, BUT…
  - CWD in more areas = increased opportunities for human exposure
  - An animal prion disease has caused disease in humans before (BSE of cattle → vCJD of humans)
  - Some animal studies suggest potential for CWD transmission to humans
  - AND multiple CWD strains exist with different transmission properties

❖ **Continued vigilance regarding this animal prion disease is essential.**

# 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**
    - To date, the number of identified human prion disease cases in these two states has been within the expected range
  - **Follow-up of individuals who consumed venison from CWD-positive deer in Wisconsin**
    - To date, no matches found among potentially exposed persons who were cross-checked with Wisconsin human prion disease surveillance data, NPDPSC data, 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

# COVID-19 and Prion Disease: Considerations

- The vast majority of US adults have received at least 1 vaccine shot. The percentage is even higher among older adults, so CJD **WILL** be diagnosed among vaccine recipients, sometimes in close proximity to the shot.
- Prion diseases are characterized by long incubation periods (typically years).
- The number of prion disease cases in the United States in 2020 (and 2021, under review) was similar to the number in 2019.
- No unusual neuropathological features have been observed at NPDPSC among cases reported as being possibly vaccine-related.

# 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, NPDPSC, and the CJD Foundation is essential.
- Improvements in pre-mortem diagnostic testing (i.e., RT-QuIC) should benefit surveillance efforts; however, autopsy remains important, and additional information is needed from a subset of RT-QuIC-positive cases.

# 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**
  - **www.cjdsurveillance.com**

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# Questions?

For more information, contact CDC
1-800-CDC-INFO (232-4636)
TTY: 1-888-232-6348 www.cdc.gov

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.