The 2026 Centers for Disease Control and Prevention (CDC) Report

Ryan Maddox, PhD

July 12, 2026

CJD Foundation Family Conference

U.S. Centers for Disease Control and Prevention

- **Nation's health protection agency**
- **Works to protect America from:**
  - Health threats
  - Safety threats
  - Security threats
- **Conducts critical science and provides health information**

"CDC serves the American public—individuals, families, and communities—who rely on accurate data, health guidance, and preventive measures."

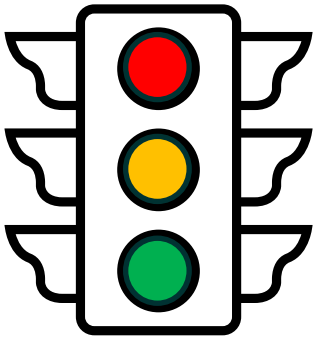
CDC Prion Team (“small but mighty”)

- Dr. Larry Schonberger: Retired, September 2025

- Dr. Joe Abrams: Epidemiologist

- Dana Haberling: Statistician

- Marissa (Person) Todd: Statistician



Prion Team Mission

- **Surveillance (for <u>human</u> prion disease) = monitoring of disease in population**
  - Estimation of prion disease rates
  - Detection of changes in disease trends over time
  - Investigation of cases of interest (identify acquired/novel forms)
  - Dissemination of subject matter expertise (e.g., infection control concerns)

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 (can be amended, process varies by state)
    - 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

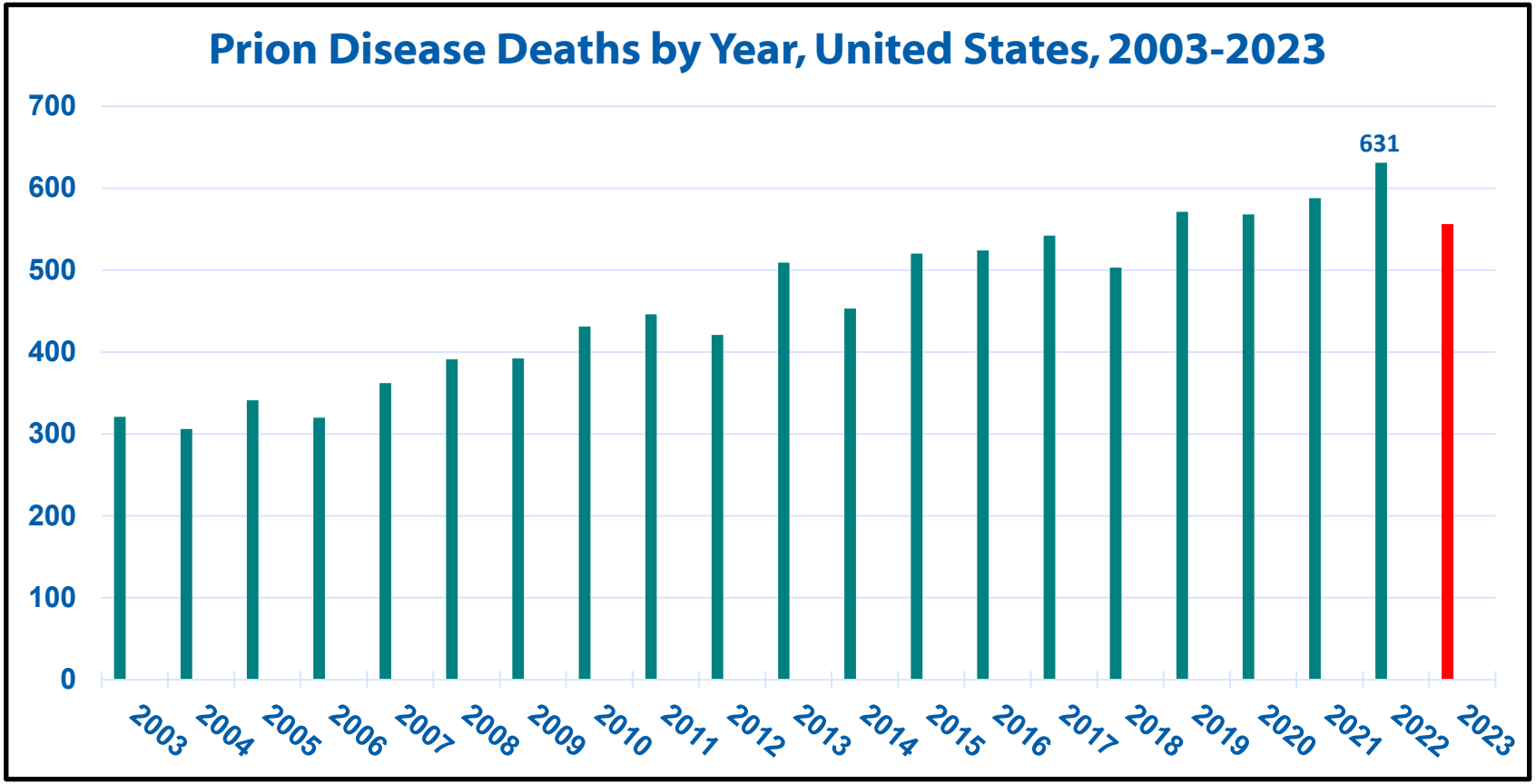
- In recent years , there have been definite or highly probable matches for >80% of cases in the national death certificate data when compared to diagnostic testing data from NPDPSC.
  - **This high percentage is a testament to NPDPSC's expertise and the increased awareness of its services.**
  - **Another lab (Mayo) now also conducts RT-QuIC testing – results are shared with NPDPSC**

Surveillance: Prion Diseases

- 9140 decedents were identified as having prion disease during 2003-2022 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, 1.2 per million.
  - Incidence among those ≥65: 6.2 cases per million per year
- Incidence (1.2/million) is likely an underestimate.
  - An RT-QuIC-positive test result strongly indicates prion disease.*
  - Review of RT-QuIC-positive cases without year of death in the NPDPSC database suggests additional prion disease cases each year.
  - Another way of looking at it: **1 CJD death for approximately every 6000 deaths overall in the US**

2003-2022? But it's 2026!

- Mortality data typically run a couple of years behind; 2024 data are delayed this year because of issues related to race categorization.
- 2023 data are available at CDC and are currently being matched to NPDPSC data.
- **This is not a dangerous gap in our surveillance**
  - We regularly review up-to-date NPDPSC data.
  - NPDPSC alerts us to young and/or unusual cases.
  - We maintain close relationships with state partners, CJD Foundation, etc.


Prion Disease Deaths by Year, United States, 2003-2023
700
600
500
400
300
200
100
0
631
2003
2004
2005
2006
2007
2008
2009
2010
2011
2012
2013
2014
2015
2016
2017
2018
2019
2020
2021
2022
2023
Death certificate data only

Surveillance: Young Cases (<30 Years)

- Most young cases have a genetic mutation or other risk factor (human growth hormone, variant CJD)
- **20 young cases 2003-2025**
  - Only 8 were sporadic forms of prion disease.
  - The remaining 12 cases were familial (5 GSS, 2 FFI, 2 fCJD), vCJD (2), and iatrogenic CJD (1, dura mater-associated).
  - **No cases diagnosed by NPDPSC in 2024 and 2025.**

frontiers | Frontiers in Cellular Neuroscience

Case Report

OPEN ACCESS

Case report: Atypical young case of MV1 Creutzfeldt-Jakob disease with unusually long survival

Lucie Yeongran Ahn, Mark L. Cohen, Ignazio Cali, Tia Russell, Jessica Ludwig, Xun Jia, Alberto Bizzi, Lawrence B. Schonberger, Ryan A. Maddox, Rohini Paul, Tania C. Ghazarian, Jaspreet Garcha, Mostafa Hammoudi and Brian Stephen Appleby

Surveillance: States

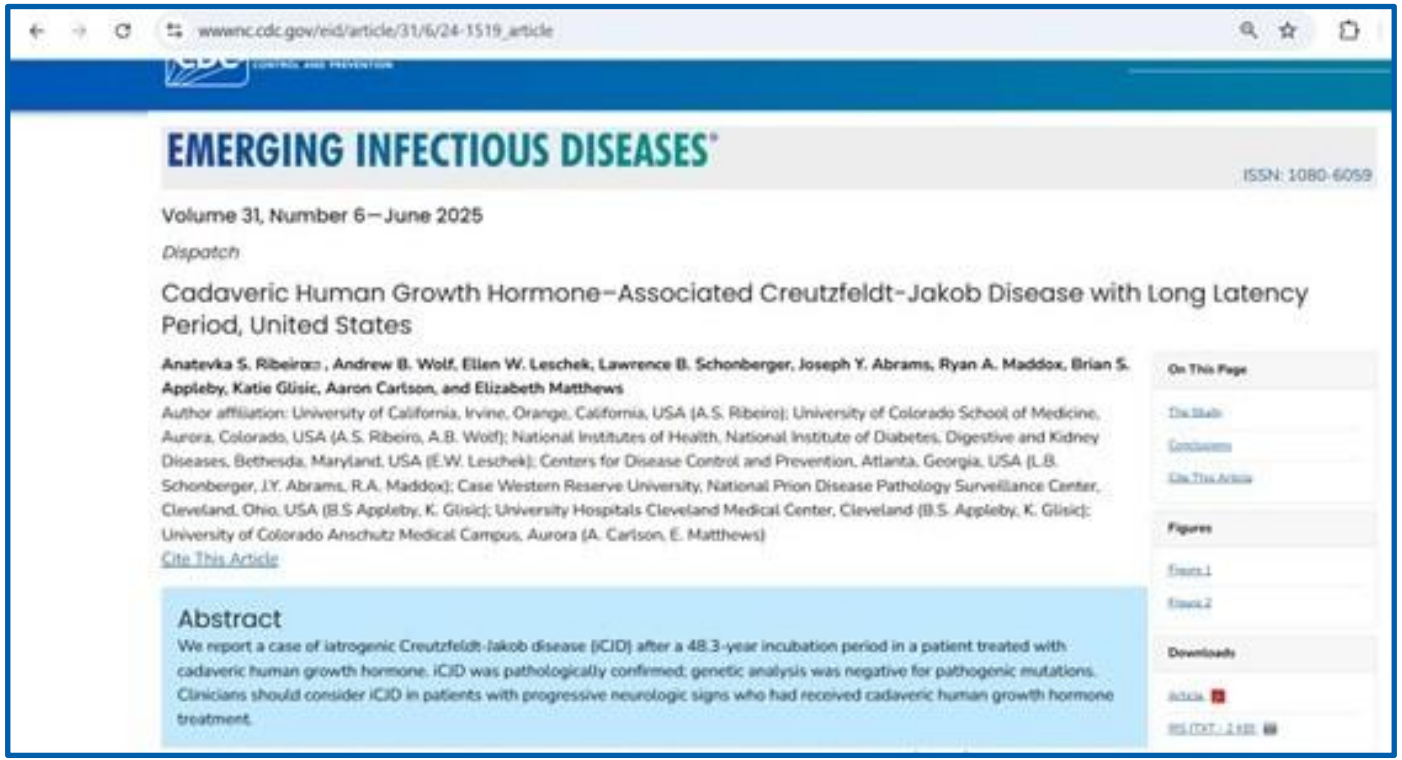
- CJD is reportable to some degree in most states.
- State reporting requirements do not necessarily translate into more accurate surveillance.
  - **Misdiagnosed case will still be a misdiagnosed case.**
- CDC helped co-author a Council of State and Territorial Epidemiologists (CSTE) position statement 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

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: Acquired Prion Disease



wwwnc.cdc.gov/eid/article/31/6/24-1519_article

EMERGING INFECTIOUS DISEASES®

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Dispatch

Cadaveric Human Growth Hormone–Associated Creutzfeldt-Jakob Disease with Long Latency Period, United States

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Cite This Article

Abstract

We report a case of iatrogenic Creutzfeldt-Jakob disease (iCJD) after a 48.3-year incubation period in a patient treated with cadaveric human growth hormone. iCJD was pathologically confirmed; genetic analysis was negative for pathogenic mutations. Clinicians should consider iCJD in patients with progressive neurologic signs who had received cadaveric human growth hormone treatment.

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Acquired Prion Disease: 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
    - **<u>NO</u> cases reported since 2021**

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

- **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; some concerns still exist about additional secondary spread
- **Updated guidance (5/2022):**
  - Removes donor deferral for geographic risk of BSE exposure
    - 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

- **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

- “The study suggests that [classic] CJD may not be transfusion-transmissible, a position in agreement with similar findings from two similar European reports…These studies have supported the conclusion that the risk, if any, of transmission of CJD by blood products is extremely small and remains theoretical.”

Funeral Homes

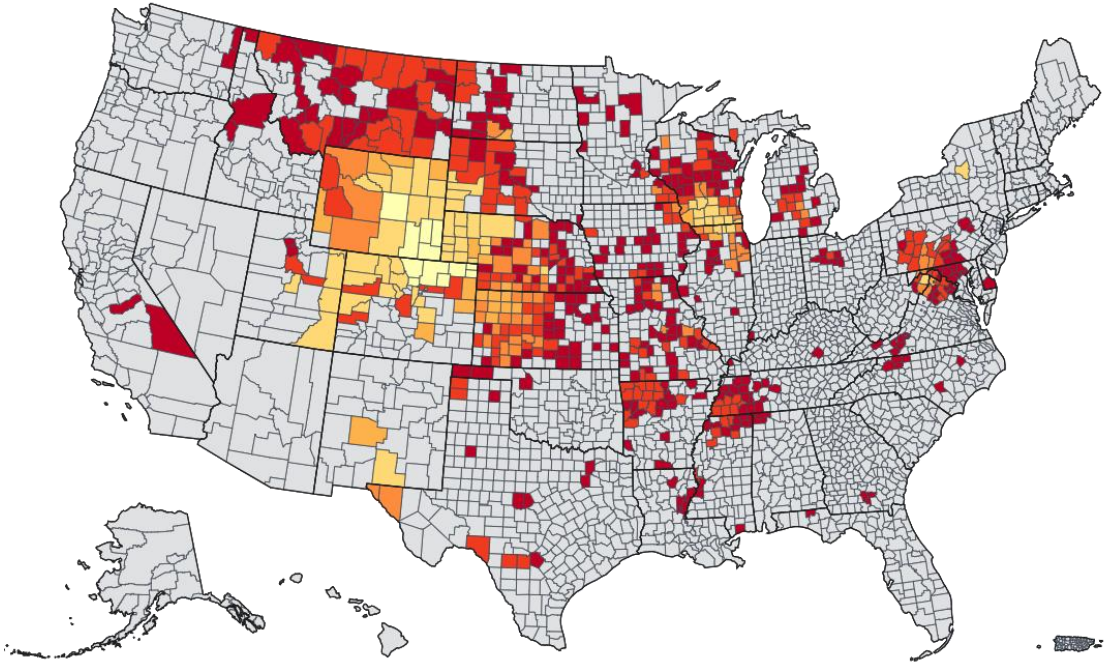
- Embalming bodies of CJD patients who have been autopsied can be safely performed, BUT a funeral home cannot be forced to accept a body.
- Education of funeral directors is important; however, some will be more open to revising policies than others.
- Information for funeral and crematory practitioners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

https://www.cdc.gov/creutzfeldt-jakob/hcp/funeral-directors/index.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 37 states (most recently Delaware) and 4 Canadian provinces**
- Also reported among free-ranging moose and/or reindeer in Norway (2016), Finland (2018), and Sweden (2019)

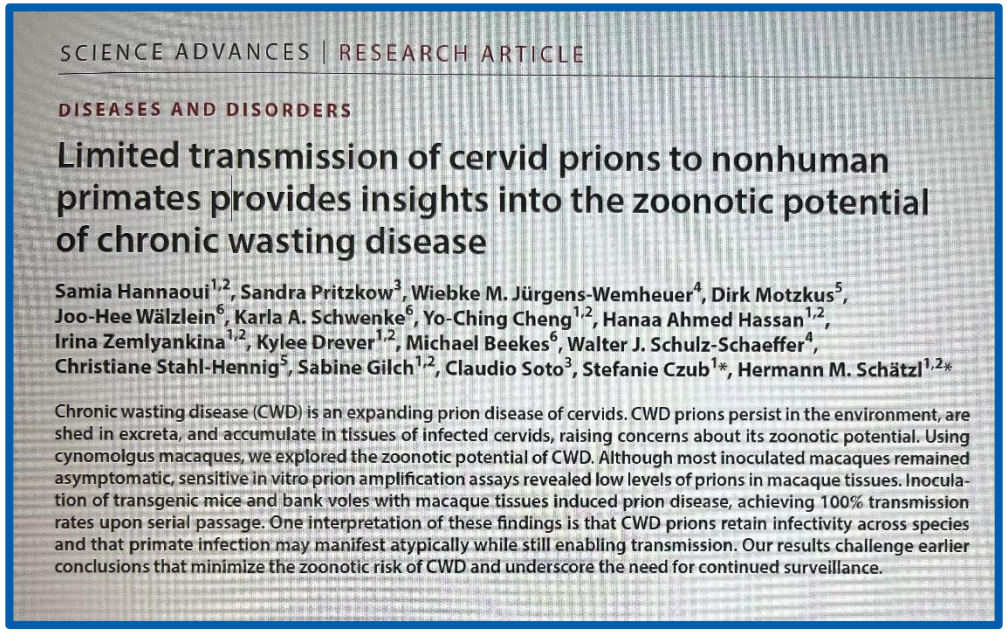
CWD: United States, May 2026



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

CWD Studies



SCIENCE ADVANCES | RESEARCH ARTICLE

DISEASES AND DISORDERS

Limited transmission of cervid prions to nonhuman primates provides insights into the zoonotic potential of chronic wasting disease

Samia Hannaoui[1,2], Sandra Pritzkow[3], Wiebke M. Jürgens-Wemheuer[4], Dirk Motzkus[5], Joo-Hee Wälzlein[6], Karla A. Schwenke[6], Yo-Ching Cheng[1,2], Hanaa Ahmed Hassan[1,2], Irina Zemlyankina[1,2], Kylee Drever[1,2], Michael Beekes[6], Walter J. Schulz-Schaeffer[4], Christiane Stahl-Hennig[5], Sabine Gilch[1,2], Claudio Soto[3], Stefanie Czub[1]*, Hermann M. Schätzl[1,2]*

Chronic wasting disease (CWD) is an expanding prion disease of cervids. CWD prions persist in the environment, are shed in excreta, and accumulate in tissues of infected cervids, raising concerns about its zoonotic potential. Using cynomolgus macaques, we explored the zoonotic potential of CWD. Although most inoculated macaques remained asymptomatic, sensitive in vitro prion amplification assays revealed low levels of prions in macaque tissues. Inoculation of transgenic mice and bank voles with macaque tissues induced prion disease, achieving 100% transmission rates upon serial passage. One interpretation of these findings is that CWD prions retain infectivity across species and that primate infection may manifest atypically while still enabling transmission. Our results challenge earlier conclusions that minimize the zoonotic risk of CWD and underscore the need for continued surveillance.

- **Continued vigilance (e.g., CDC/state studies) regarding CWD essential**

Colorado and Wyoming: Hunter registry data

- **Follow-up of persons who hunted in Colorado and Wyoming, where CWD has been present for years, and identifying those who died of prion disease**
- **Hunter registry data from Colorado Parks and Wildlife (CPW) and Wyoming Game and Fish Department (WGFD) since mid-1990s**
  - CPW: 1,194,956 hunters
  - WGFD: 556,758 hunters
- **Hunter data were cross-checked with mortality data to determine mortality status and cause of death to identify prion disease cases.**
  - State mortality records
  - National Death Index (National Center for Health Statistics)

Colorado and Wyoming	CJD deaths through mortality matching	
	Observed	Expected (95% CI)
Colorado		
All hunters	**26**	**23.3 (14 – 33)**
Hunters who hunted in areas known to be CWD-endemic or purchased all-state license	**25**	**22.3 (14 – 32)**
Hunters who hunted in areas known to be CWD-endemic	**19**	**13.4 (7 – 21)**
Wyoming		
All hunters	**8**	**5.2 (1 – 10)**
Hunters who hunted in areas known to be CWD-endemic or purchased all-state license	**5**	**2.5 (0 – 6)**
Hunters who hunted in areas known to be CWD-endemic	**4**	**1.6 (0 – 5)**

- To date, the number of identified human prion disease cases in these two states has been within the expected range.

Wisconsin: Follow-up of individuals who consumed venison from CWD-positive deer in Wisconsin

Wisconsin		
	Database	Matches*
Hunters who consumed venison from CWD+ deer with Date of Birth data (2003-2023)	637	0
Consumed venison from CWD+ deer with Date of Birth data AND processed their own animal	303	0
Hunters who harvested a positive deer (2020-2024)	4589	0

* NPDPSC data, national mortality data, state mortality data

- To date, no matches have been 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.

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).
- No unusual neuropathological features have been observed at NPDPSC among cases reported as being possibly vaccine-related.
- The numbers of prion disease cases in the United States in 2019, 2020, and 2021 were very similar.

Conclusion

- 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.

Acknowledgements

- All the wonderful people at:
  - **CJD Foundation**
  - **NPDPSC**
  - **State and local public health departments**

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.