

The 2020 Centers for Disease Control and Prevention Report

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CJD Foundation Webinar

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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 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

- ❑ 6781 decedents were identified as having prion disease during 2003-2018 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).
- ❑ 14 cases were <30 years of age: 7 cases *per billion* per year.
 - Only 3 were sporadic forms of prion disease, 1 sporadic CJD and 2 sporadic fatal insomnia.
 - The remaining 11 cases were familial (5 GSS, 2 FFI, 1 CJD), variant CJD (2), and iatrogenic CJD (1, dura mater-associated).
- ❑ Incidence among those ≥ 65 : 6 cases per million per year.
- ❑ 1 CJD death for approximately every 6,000 deaths overall in the US each year

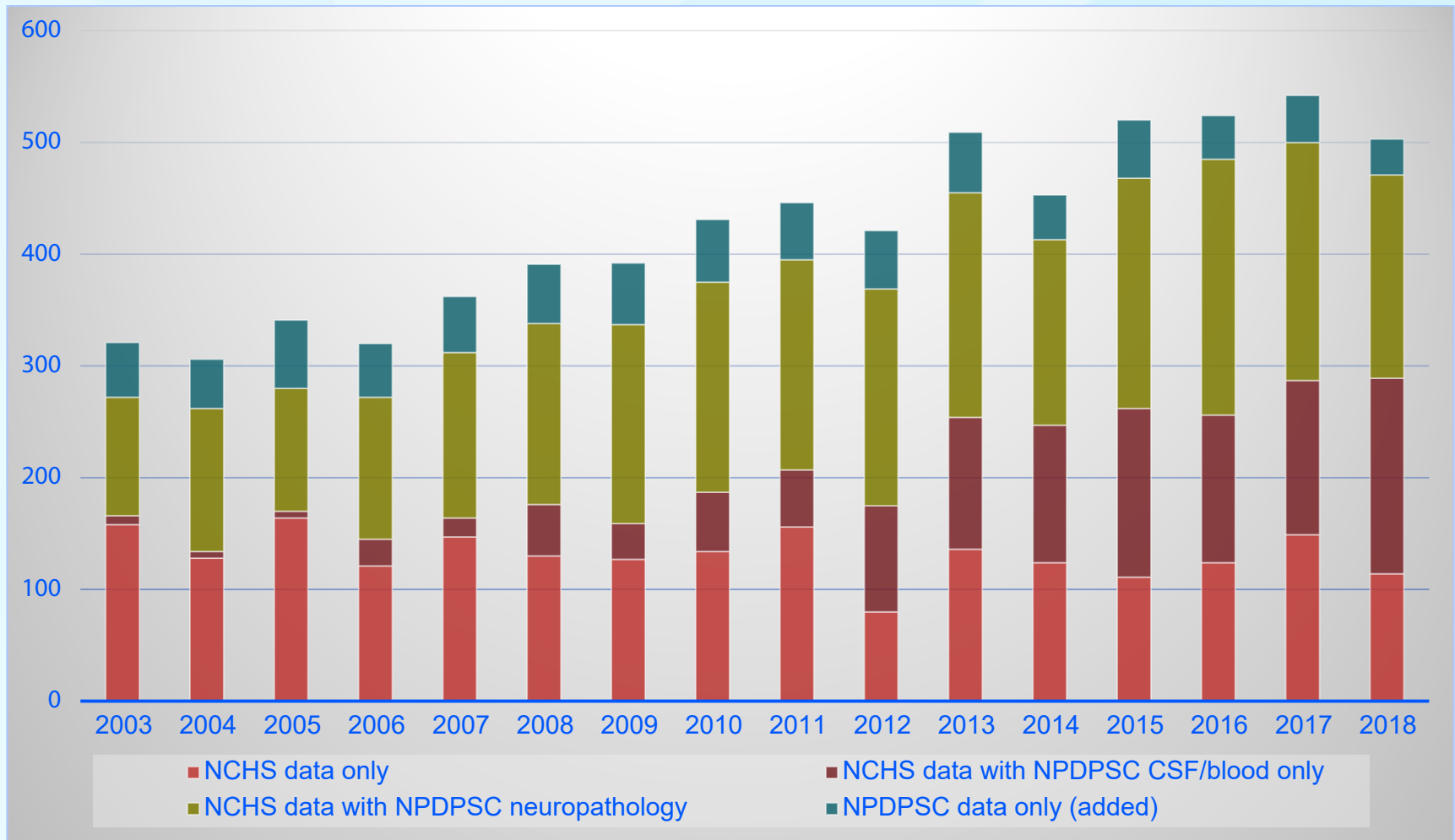
Surveillance: Prion Diseases

- ❑ Of the total cases each year during 2003 -2018, 6%-18% were the result of additions to the death certificate data based on positive neuropathologic and/or genetic findings.
- ❑ Out of all the death certificates with prion disease indicated, 2% - 6% were removed annually based on negative neuropathology results.
 - NPDPS'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: Prion Diseases

- ❑ 2017 data (542 cases):
 - 10 (2%) autopsy-negative cases excluded out of 511 cases identified through death certificate data
 - 23/121 (19%) death certificate cases with RT-QulC and no autopsy had a negative RT-QulC
 - 42 autopsy-positive NPDPSC cases added (8% of total)
- ❑ 2018 data (503 cases):
 - 8 (2%) autopsy-negative cases excluded out of 479 cases identified through death certificate data
 - 21/157 (13%) death certificate cases with RT-QulC and no autopsy had a negative RT-QulC
 - 32 autopsy-positive NPDPSC cases added (6% of total)

Prion disease cases by year, United States, 2003-2018



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.

Surveillance: Variant CJD



- ❑ Variant CJD is the human form of bovine spongiform encephalopathy (BSE, or “mad cow disease”).
- ❑ 232 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.

Tissue Donation

- ❑ FDA Guidance for Industry: 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 Donation



- ❑ 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>

Blood Donation: New Recommendations (4/2020)

❑ FDA Guidance for Industry: Blood

Ineligible:

- Persons with a history of transfusion in U.K. (i.e., England, Northern Ireland, Scotland, the Isle of Man, the Channel Islands, Gibraltar, or the Falkland Islands), France, or Ireland from the beginning of 1980 to present.
- Persons who have spent three months or more cumulatively in the U.K. (i.e., England, Northern Ireland, Scotland, Wales, the Isle of Man, the Channel Islands, Gibraltar, or the Falkland Islands) from 1980 to 1996.
- Persons who have spent 5 or more years cumulatively in France or Ireland from the beginning of 1980 to the end of 2001.
- Persons who have been diagnosed with vCJD, CJD, or any other transmissible spongiform encephalopathy or who have a blood relative diagnosed with genetic CJD (e.g., fCJD, GSS, or FFI).

Blood Donation: New Recommendations (4/2020)

❑ Updated guidance:

- Drops the recommendation to defer former or current U.S. military personnel, civilian military personnel and their dependents who resided at U.S. military bases.
- Drops the recommendation to defer potential donors who spent 5 years or more in Europe from 1980 to the present, with the exceptions listed previously.
- No longer recommends questioning donors regarding blood relatives with genetic CJD; donors previously deferred for having a blood relative with CJD can be reentered if the blood relative was not diagnosed with a genetic form of the disease.

Blood Donation: New Recommendations (4/2020)

- Note that for eligible potential donors who have been permanently deferred under previous criteria, it is our understanding that there will be a process for removing that deferral, although we do not know when it will be made available.

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; this risk remains theoretical.”
 - United Kingdom 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 in a few cases.

Funeral Homes

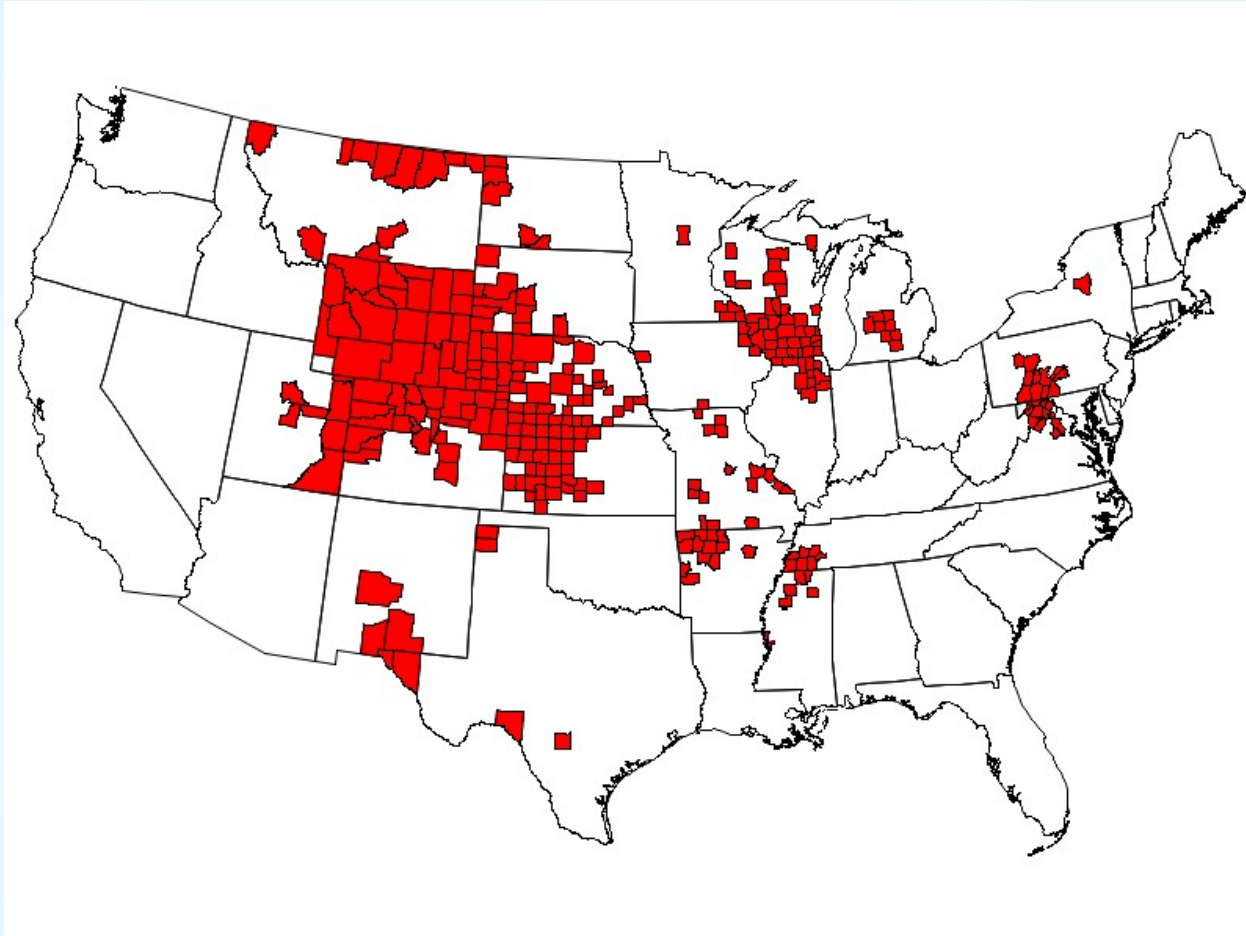
- ❑ Embalming bodies of CJD patients who have been autopsied can be safely performed, BUT a funeral home cannot be forced to accept a body.
- ❑ Education of funeral directors is important; however, some will be more open to revising policies than others.
- ❑ Information for funeral and crematory practitioners 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, January 2020



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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5:51

ANIMALS

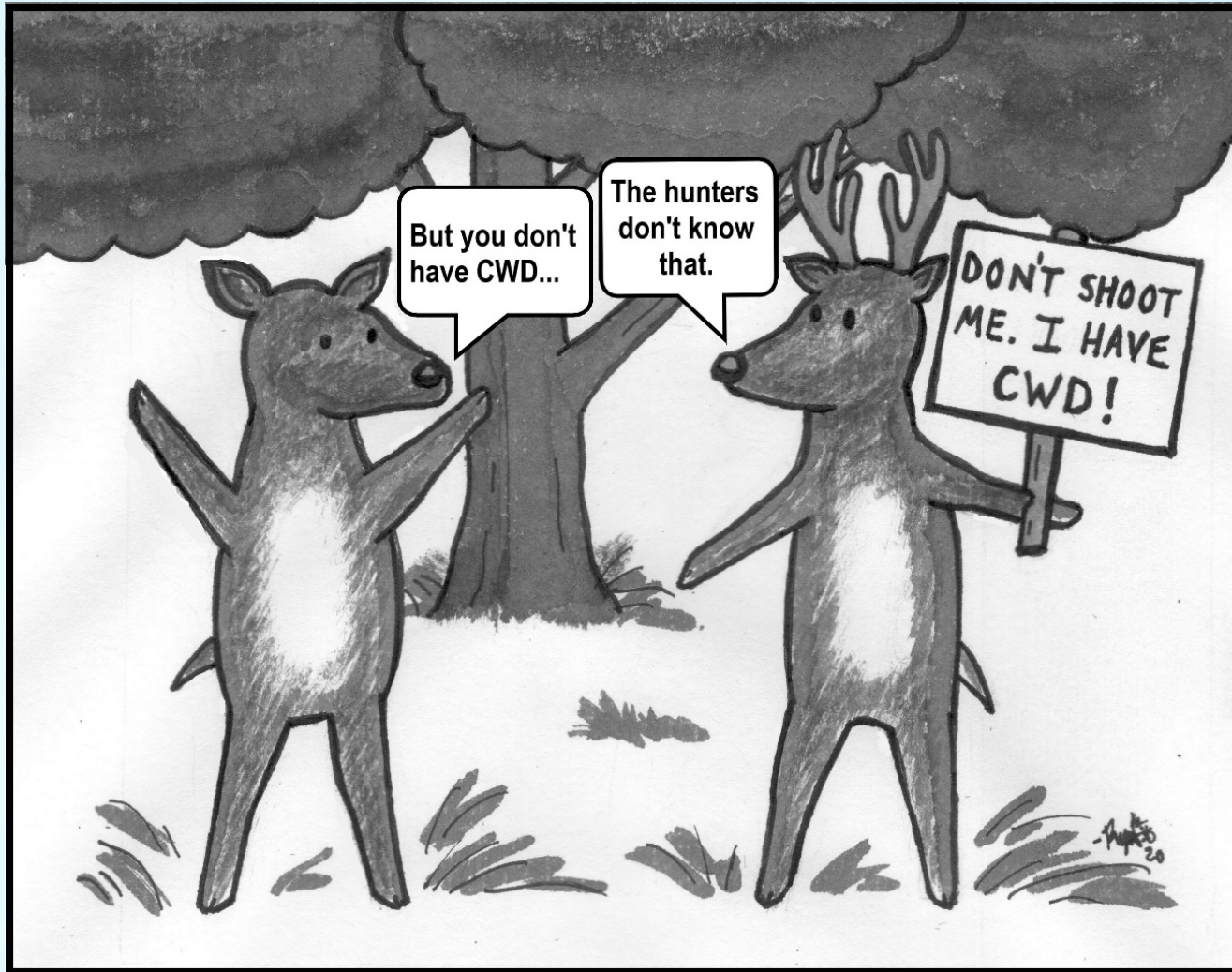
The Dangers Of 'Zombie Deer Disease'

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

- ❑ Before COVID-19, there was ZOMBIE DEER DISEASE.
- ❑ To date, there is no strong epidemiologic evidence for the occurrence of CWD in people.

Oh Deer...

- ❑ There is no strong 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)
 - AND some animal studies suggest potential for CWD transmission to humans



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
 - To date, no matches found among potentially exposed persons who were cross-checked 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, 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.

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

Questions?

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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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