# The Centers for Disease Control and Prevention Report: Prion Disease Activities at CDC

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Division of High-Consequence Pathogens and Pathology

# Objective

- **To describe prion disease activities of the Centers for Disease Control and Prevention (CDC)**
  - Surveillance
  - Investigation
  - Consultation


# Surveillance

# What is surveillance?

- **Surveillance – monitoring of disease in population**
  - Estimation of prion disease rates
  - Detection of changes in epidemiology of disease over time
  - Monitoring of possible occurrence of variant CJD or novel prion diseases
  - Gaining of knowledge about prion diseases

# Surveillance

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

# CJD Surveillance Difficulties

- Lack of reliable laboratory diagnostic test prior to patient death
- Disease confirmed only by pathology
- US autopsy rates historically low
- Common-source cases: long incubation period makes tracking or identifying "source" difficult

# Surveillance mechanisms

- Periodic review of national cause-of-death data
- Ongoing review of clinical and pathologic records of CJD decedents aged <55 years
- Collaboration with the National Prion Disease Pathology Surveillance Center (NPDPSC)
- Assessment of potential cases (iatrogenic, vCJD, etc.) reported by the media, the public, clinicians, and public health departments
- Collaborative surveillance of special groups

# National cause-of-death 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 easily obtainable, ongoing

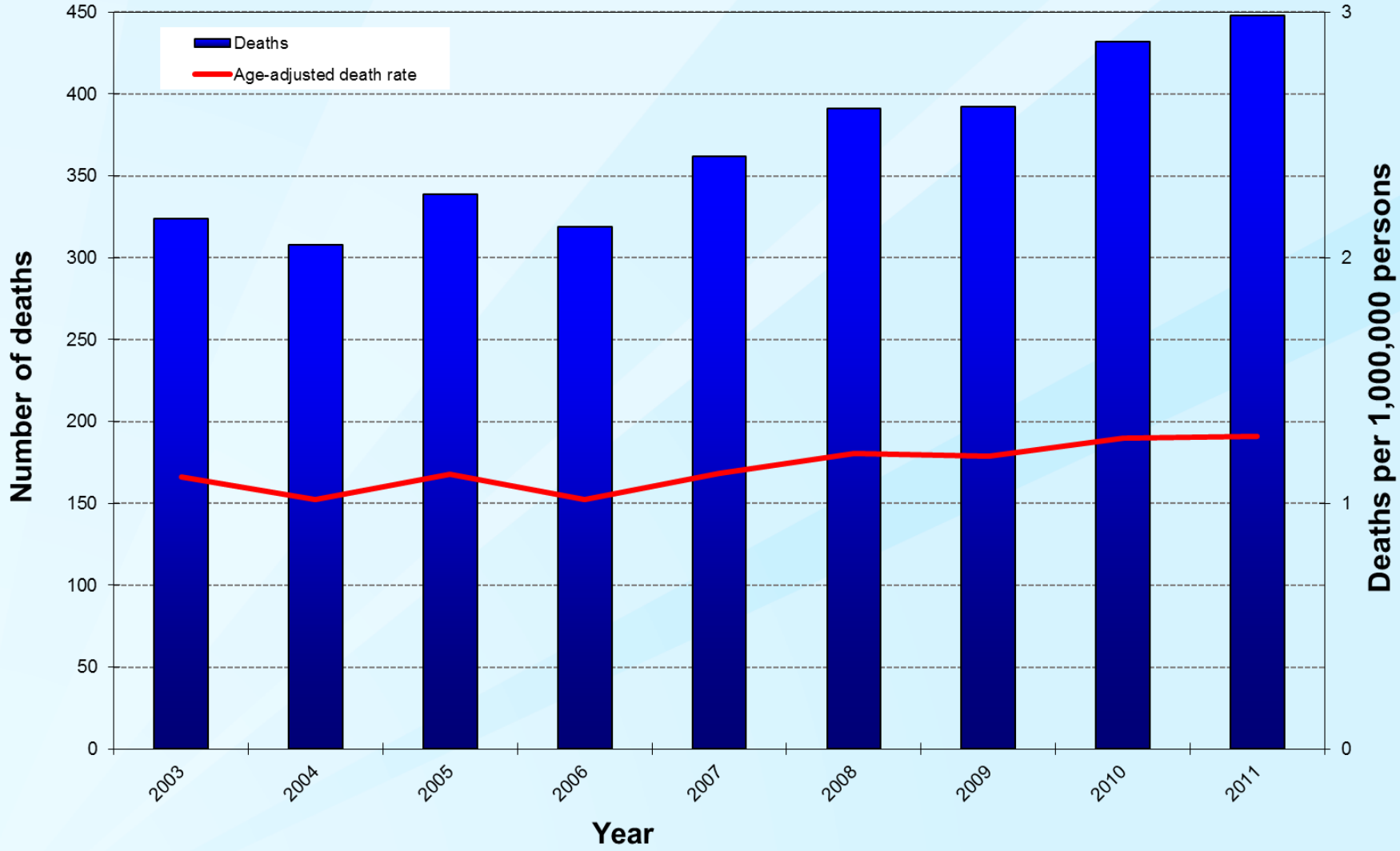
# Collaboration with NPDPSC

- **Neuropathology necessary for diagnosis confirmation**
- **NPDPSC collaboration is a valuable surveillance tool for some states.**
  - First notification of case may be NPDPSC report
- **Sentinel for unique prion disease cases**

# Collaboration with NPDPSC

- **To more accurately determine prion disease incidence in the United States, NCHS death certificate data are being adjusted through a matching process with NPDPSC data.**
  - Based on NPDPSC neuropathology results, cases are added to or subtracted from NCHS data.
- **For 2003-2011, a majority (56.5%) of the NCHS decedents had a corresponding match in the NPDPSC database, indicating that the center had some knowledge of the case (e.g., brain, blood, csf specimen). Almost half (49.0%) of the NCHS decedents 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.**

# Figure 2: Creutzfeldt-Jakob disease deaths and age-adjusted death rate, United States, 2003-2011*




* Deaths obtained from NCHS multiple cause-of-death data and NPDPSC data; multiple cause-of-death data are based on ICD-10 codes with available computerized literal death certificate data. Deaths include familial prion disease. Rates are adjusted to the US standard 2000 projected population.

# Collaboration with states

- **CDC collaborates with several states on surveillance projects.**
- **Goals:**
  - Develop ways of enhancing surveillance
  - Identify barriers to surveillance and find solutions
    - Resistance by pathologists to perform autopsies
    - Unfamiliarity of clinicians with prion diseases
    - Concerns regarding infection control risks

# Surveillance of special groups

- Collaborative surveillance of special groups to ascertain additional information on prion disease transmission properties

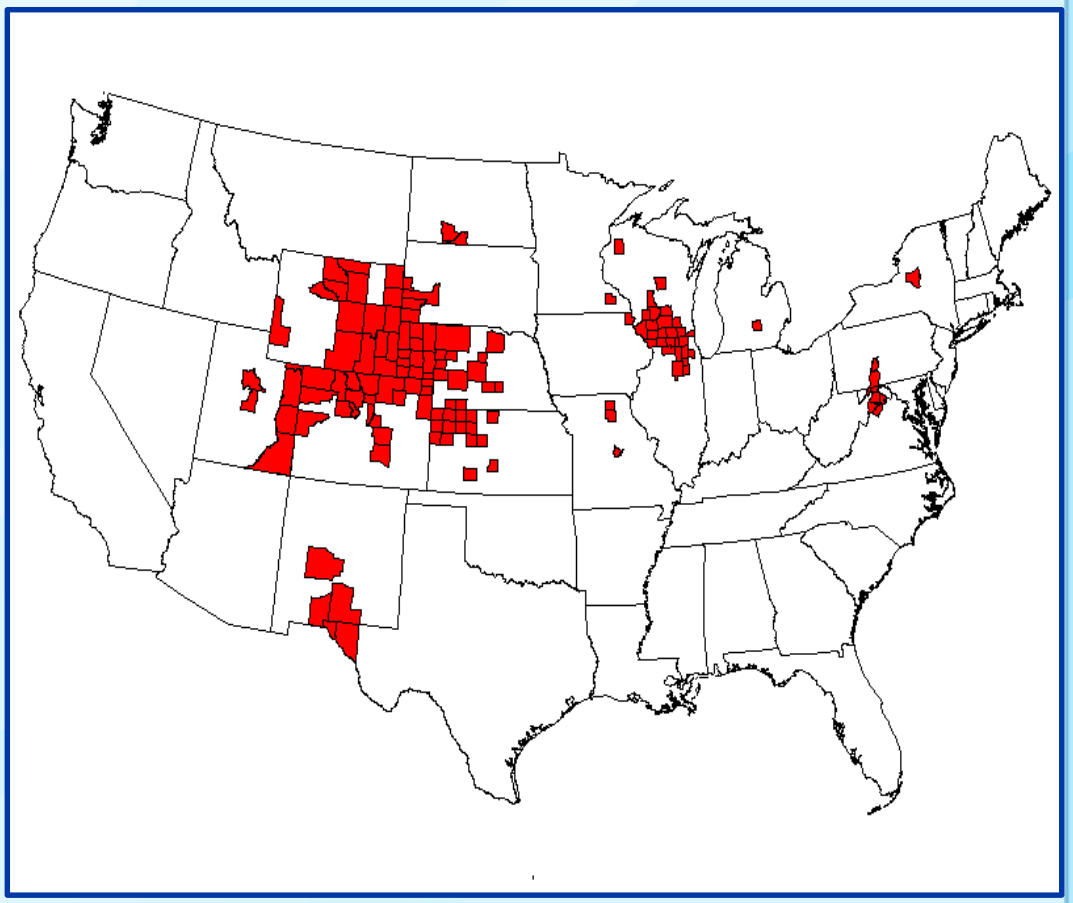
# Blood study

- Goal: To determine whether CJD is transmissible through blood
- Study: Follow-up of recipients of blood components from donors who subsequently developed CJD
- Results: No evidence of CJD transmission through blood to date

# Hunter studies

- **Goal: To determine whether chronic wasting disease (CWD), a prion disease of deer and elk, can cause disease in humans**

- **Studies:**
  - **Follow-up of persons who hunted in Wyoming and Colorado, where CWD is found, and identifying those who died of prion disease**
  - **Follow-up of hunters who consumed venison from CWD-positive deer in Wisconsin**



**Chronic Wasting Disease Among Free-Ranging Cervids by County, United States, June 2015**

# Hunter studies

- **Wyoming: 2.2 million records of licenses purchased during 1996-2013, representing about 0.6 million hunters.**
  - 4 Wyoming hunters have been identified through mortality matching to have died of CJD (expected: 4.63 cases, 95% CI = 1-9).
- **Colorado: 6.1 million records of licenses purchased during 1995-2011, representing about 1.1 million hunters.**
  - 11 Colorado hunters have been identified through mortality matching to have died of CJD (expected: 10.19 cases, 95% CI = 4-17).
- **Wisconsin: Approximately 400 hunters identified as having consumed venison from CWD-positive deer.**
- **Results: Prion disease cases among these groups within expected range so far, but many years of follow-up necessary**

# The Future

- **The percentage of the US population ≥65 years of age is projected to increase from 13.0% in 2010 to 20.3% in 2030.**
- **Applying 2008-2010 CJD incidence rates to US census projections, in 2030 there may be 460 CJD decedents ≥65 years of age in the United States.**
  - This number would represent an 85% increase compared to the 2008-2010 average of 248 cases for this age group.
- **It is important that medical personnel are educated about the disease and familiar with recommended infection control guidelines to minimize undue concerns and risks related to transmission.**

**Investigation**

# Investigation

- **CDC works with states to investigate cases of concern:**
  - Possible clusters
  - Iatrogenic cases
  - Variant CJD cases

MMWR
Morbidity and Mortality Weekly Report
Weekly May 14, 2004 / Vol. 53 / No. 18

# Creutzfeldt-Jakob Disease Not Related to a Common Venue — New Jersey, 1995–2004

*On May 7, this report was posted as an* MMWR Early Release *on the* MMWR *website* (http://www.cdc.gov/mmwr).

Beginning in June 2003, the New Jersey Department of Health and Senior Services (NJDHSS) and CDC were notified of a suspected cluster of deaths caused by Creutzfeldt-Jakob disease (CJD) in persons reportedly linked to Garden State Racetrack in Cherry Hill, New Jersey. Concerns were raised that these deaths might have resulted from consumption of meat contaminated with the agent causing bovine spongiform encephalopathy (BSE, commonly called "mad cow disease") served at racetrack restaurants during 1988–1992.

# Cluster investigations

- **When investigating a cluster, we must keep in mind:**
  - Are all cases actually CJD?
  - Do some or all the CJD cases have a genetic component?
  - How large of an area is under consideration (e.g., a hospital may serve 1 county or 3 states)?
  - How long have cases resided in the area?

# Investigation of dura mater case, 2007

## A Case of Creutzfeldt-Jakob Disease Associated With a Dura Mater Graft in the United States

David B. Blossom, MD, MS; Ryan A. Maddox, MPH;
Suzanne F. Beavers, MD; Kelly A. Church, MD;
Doug A. Thoroughman, PhD;
Lawrence B. Schonberger, MD, MPH;
Ermias D. Belay, MD

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We describe a case of Creutzfeldt-Jakob disease associated with a dura mater graft (Lyodura brand) in a 26-year-old man who underwent several neurosurgical procedures as a child. Clinicians and infection control personnel should be aware that recipients of Lyodura brand dura mater grafts processed before May 1987 may remain at increased risk for Creutzfeldt-Jakob disease throughout their lives.

*Infect Control Hosp Epidemiol* 2007; 28:1396-1397

# Iatrogenic cases

- **Iatrogenic CJD cases will continue to occur.**
  - Dura mater, hGH, and corneal transplant-associated cases may have been exposed years ago, but long incubation period makes additional CJD cases possible.
  - Abrams, et al.: Lower risk of Creutzfeldt-Jakob disease in pituitary growth hormone recipients initiating treatment after 1977
- **Neurosurgical instrument-related cases appear to be almost non-existent; however, case investigation can be difficult.**

# Investigation of a variant CJD case in the United States, 2014

## 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.

# Variant CJD is transmissible through blood

- **Bloodborne transmission of vCJD is an issue of concern.**
- **Transmission of vCJD through blood has been reported in the United Kingdom.**
  - Donors developed vCJD symptoms months or even years after donation
- **Deferral policy for blood donors in the United States with extended travel to the United Kingdom and Europe**
  - Policy currently under review by FDA

**Consultation**

# Consultation

- CDC provides prion disease information, references, and recommendations on its website:
  - http://www.cdc.gov/prions/cjd/index.html
- We are available to give advice by phone or e-mail.
- CJD Foundation is a valuable resource for family members of patients.

# Consultation – common topics

- Hospital infection control issues
- Media report clarification
- Funeral home procedures
- Caregiver concerns

# Conclusion

- 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.

# Conclusion

- 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
- **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/npdpsc/

# Acknowledgments


- **CDC**
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  - Dr. Jim Sejvar
  - Ms. Teresa Hammett
  - Mr. Joe Abrams

- **All the wonderful people at:**
  - 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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