

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

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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 CJD rates
 - Detection of changes in epidemiology of disease over time
 - Monitoring of possible occurrence of variant CJD
 - Gaining of knowledge about CJD

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**

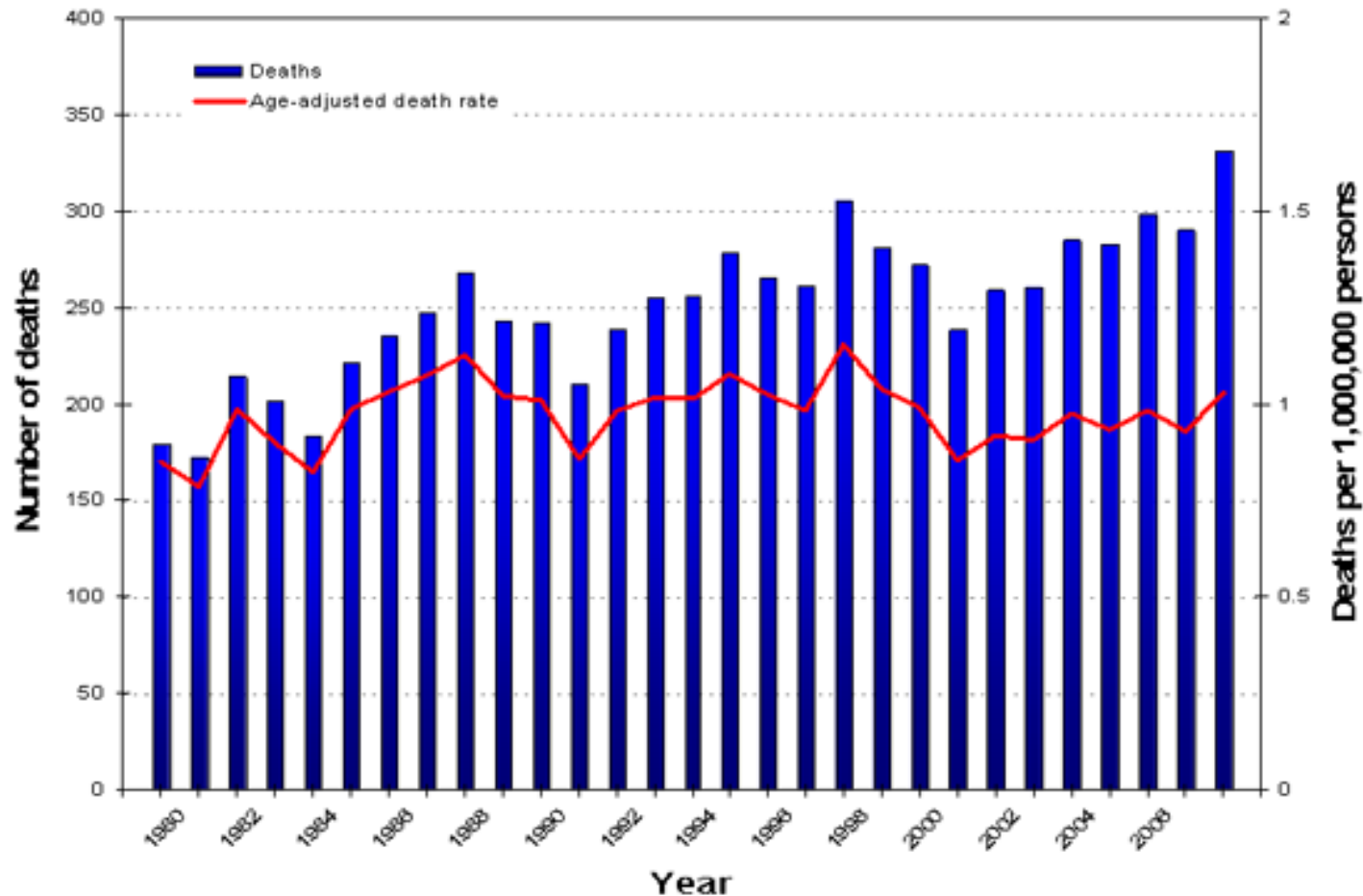
Surveillance mechanisms

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

Creutzfeldt-Jakob disease deaths and age-adjusted death rate, United States, 1979-2007*



*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; includes 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 disease
 - Concerns regarding infection control risks

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**

Surveillance of special groups

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

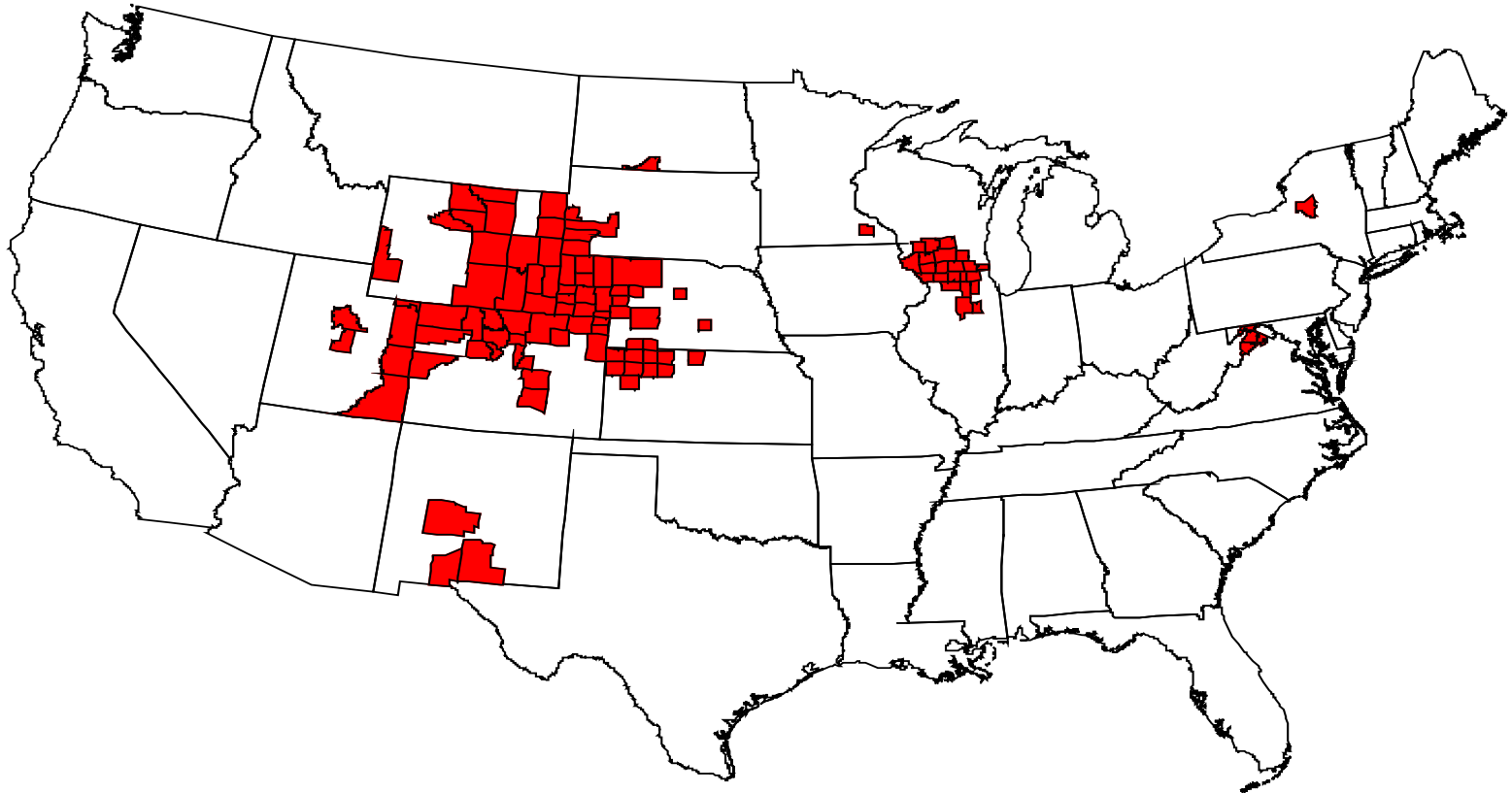
Blood study

- ❑ **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 study

- ❑ **Goal:** To determine whether chronic wasting disease (CWD), a prion disease of deer and elk, can cause disease in humans
- ❑ **Study:** Follow-up of persons who hunted in Colorado and Wyoming, where CWD is found, and identifying those who died of prion disease
- ❑ **Results:** Prion disease cases among this group within expected range so far

Chronic Wasting Disease Among Free-Ranging Cervids by County, United States, July 2011



CJD surveillance by race and ethnicity

- ❑ **During 1994-2007, 150 of 3903 (3.8%) CJD decedents reported in the US were classified as black race.**
- ❑ **The average annual age-adjusted incidence for blacks was 0.41 per million persons, compared to 1.1 for whites (RR 0.38, 95% CI 0.32-0.44).**
- ❑ **The incidence of CJD among whites is more than two and a half times that of blacks. The differences between black and white CJD decedents warrant further investigation.**

CJD surveillance by race and ethnicity

- ❑ **According to the 2010 census, there are 50.5 million Hispanics in the US, 16.3% of the total population.**
 - The US Hispanic population grew by 15.2 million between 2000–2010, a 43% increase, and this population is expected to continue to increase substantially in the coming decades.
- ❑ **During 1997–2008, 160 US CJD decedents were reported as being of Hispanic ethnicity, for an average annual age-adjusted incidence of 0.65 per million population.**
 - This incidence is significantly lower than that for US non-Hispanic CJD decedents (RR=0.6; 95% CI 0.5–0.7).

CJD surveillance by race and ethnicity

- ❑ **The lower incidence of CJD among Hispanics in the US may be at least partly due to:**
 - Underreporting of Hispanic ethnicity on death certificates relative to surveys and censuses.
 - Possible genetic differences between Hispanics and non-Hispanics.
 - Selective return migration of less healthy individuals (upon illness onset, some Hispanics may return to their country of origin for treatment, family support, etc.). This scenario has been suggested as one possible reason for the overall lower mortality rates reported among Hispanics compared to non-Hispanic whites in the US.



Investigation

Investigation

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



MMWRTM

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 E. Beavers, MD; Kelly A. Church, MD;
Doug A. Thoroughman, PhD;
Lawrence B. Schonberger, MD, MPH;
Ermias D. Belay, MD

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.
- ❑ **Neurosurgical instrument-related cases appear to be almost non-existent; however, case investigation can be difficult.**

Investigation of a variant CJD case, 2004

Variant Creutzfeldt-Jakob Disease Death, United States

Ermias D. Belay,* James J. Sejvar,* Wun-Ju Shieh,* Steven T. Wiersma,† Wen-Quan Zou,‡
Pierluigi Gambetti,‡ Stephen Hunter,* Ryan A. Maddox,* Landis Crockett,† Sherif R. Zaki,*
and Lawrence B. Schonberger*

The only variant Creutzfeldt-Jakob disease (vCJD) patient identified in the United States died in 2004, and the diagnosis was confirmed by analysis of autopsy tissue. The patient likely acquired the disease while growing up in Great Britain before immigrating to the United States in 1992. Additional vCJD patients continue to be identified outside the United Kingdom, including 2 more patients in Ireland, and 1 patient each in Japan, Portugal, Saudi Arabia, Spain and the Netherlands. The reports of bloodborne transmission of vCJD in 2 patients, 1 of whom was heterozygous for methionine and valine at polymorphic codon 129, add to the uncertainty about the future of the vCJD outbreak.

A few words on variant CJD

- ❑ Variant CJD is the human form of bovine spongiform encephalopathy (BSE, or “mad cow disease”).**
- ❑ Three cases in the United States, and two in Canada, have been identified, but none are believed to have been exposed in North America.**
- ❑ Variant CJD and classic CJD are separate diseases.**

Variant CJD is transmissible through blood

- ❑ **Bloodborne transmission of vCJD is a recent 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**



Consultation

Consultation

- ❑ **CDC provides prion disease information, references, and recommendations on its website:**
 - <http://www.cdc.gov/ncidod/dvrd/cjd/>
- ❑ **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**
- ❑ CDC website provides infection control guidelines, information for funeral directors, and facts about prion diseases**

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, public education, and physician awareness of NPDPSC's services.**

CJD resources

❑ CJD Foundation

- 1-800-659-1991
- help@cjd.foundation.org

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

- 404-639-3091
- <http://www.cdc.gov/ncidod/dvrd/cjd/index.htm>

❑ National Prion Disease Pathology Surveillance Center

- 216-368-0587
- cjdsurv@cwru.edu

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- NPDPS
- State and local public health departments

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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Division of High-Consequence Pathogens and Pathology

