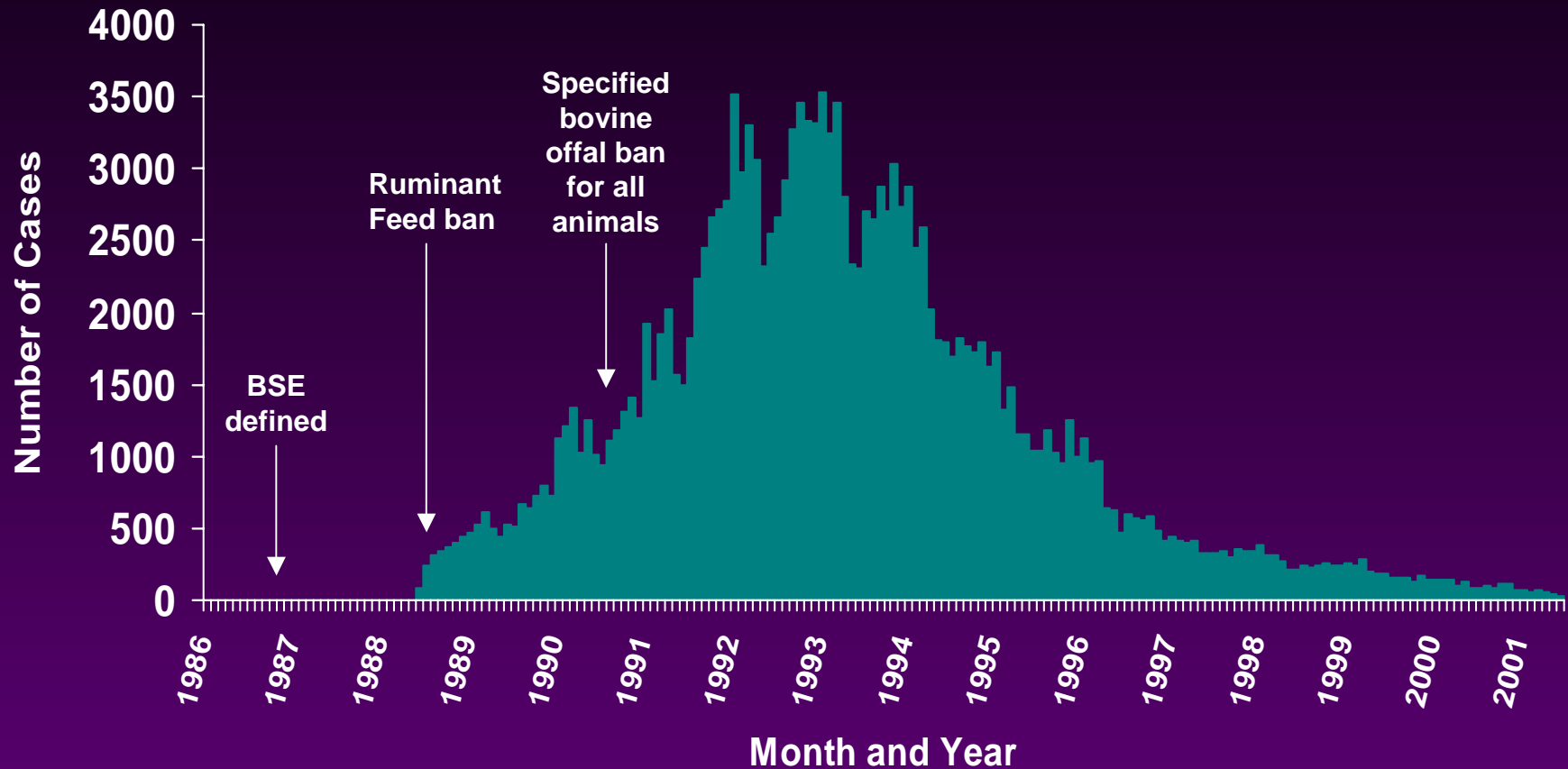




# Impact of the BSE/vCJD Outbreak: U.S. Concerns for TSEs



# Confirmed BSE Cases by Month and Year of Restriction, Great Britain, June 1988-June 2001



# BSE Impacts

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- **Transmission of the agent among cattle and to other animals.**
- **Transmission to humans causing vCJD predominantly among young patients.**
- **Invariably fatal nature of the diseases with no effective therapy.**
- **Economic impacts of BSE**
  - **During September 2001-January 2002, the farming and food industries in Japan reportedly lost \$1.5 billion because of BSE.**

# U.S. Concerns Related to TSEs

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- **Concerns related to possible introduction of BSE into the United States.**
- **Concerns related to possible occurrence of vCJD and/or its possible secondary spread via blood and blood products.**
- **Concerns related to CJD infection control issues and the possible zoonotic transmission of animal TSEs prevalent in the United States.**

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**PARDONS: BILL'S DESIGNING WOMEN • RAP: ALL ABOUT EVE**

# Newsweek

March 12, 2001 • \$3.50

The  
Slow,  
Deadly  
Spread  
of

# Mad Cow Disease

How  
It Could  
Become an  
Epidemic

[newsweek.msnbc.com](http://newsweek.msnbc.com)

**CDC**  
FEDERAL CENTERS FOR DISEASE CONTROL AND PREVENTION

# Moving to Keep the Beef Out of Disease's Shadow

*FDA, Industry Work to Bolster Mad Cow Safeguards*

By MARG KATZMAN  
Washington Post Staff Writer

In Texas, more than 1,000 cattle being fattened for slaughter were immediately quarantined after federal officials discovered the animals had been inadvertently fed the kind of bovine meal believed to have spread mad cow disease throughout Europe.

In New York City, health officials became alarmed after learning that Mamba fruit chews were being sold in stores even though the German-made candy had been banned in Poland because it was made with gelatin that included melted-down beef parts.

And in Oklahoma, a captive herd of 140 elk was segregated from other animals after several were found to have died from "chronic wasting disease," which is closely related to mad cow disease.

In each case, federal authorities eventually concluded there was no danger. But the incidents show how mad cow disease, the fatal brain disease that has caused panic over beef in Europe, is casting a shadow in the United States.

tie in the mid-1990s has got public health officials on high alert.

The European epidemic has been made worse—and more politically damaging—by misplaced official assurances that there was no problem. American officials are taking the opposite approach, explaining that an isolated case of mad cow disease may show up sooner in U.S. cattle. But they say that precautions in place for several years will keep the disease from spreading.

"Nothing in the world is risk-free, but we can say that the risk of us having a situation like in the United Kingdom with thousands of sick animals showing up is very, very small," said George Gray of the Harvard School of Public Health, the lead researcher on a two-year risk assessment of mad cow disease for the Agriculture Department. "Even if it does show up here, the protective measures in place make it extremely unlikely there will be a significant public health or animal health problem."

Because of concerns over mad cow disease, the United States prohibited imports of British animals in 1989 and European animals in 1997. The FDA stopped the practice of feeding



Trying To Keep  
**"Mad Cow Disease"**  
 Out Of U.S. Herds

By Linda Dow



Washington Post

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## FDA: Texas Cows Not a Threat

By Lauran Neergaard  
 AP Medical Writer

Tuesday, Jan. 30, 2001; 6:25 p.m. EST

WASHINGTON — About 1,200 Texas cattle ate animal feed containing ingredients banned as a precaution against mad cow disease but not enough to threaten the food supply, government investigators concluded Tuesday.

The cattle have been quarantined at a Texas feed lot since last week as the Food and Drug Administration investigated whether a Purina Mills Inc. plant violated a federal ban on feeding beef byproducts to other cows.

Mad cow disease, also known as bovine spongiform encephalopathy or BSE, has not been found in U.S. cattle. But cows can catch the illness by eating feed made from the parts of infected cows or certain other animals. The animal feed ban is a precaution to keep BSE from spreading should a U.S. cow ever become infected.

CDC

PHOTO BY AP/WIDEWORLD



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## FDA: Candy With Beef Gelatin Safe

The Associated Press  
Tuesday, Jan. 30, 2001; 9:09 p.m. EST

NEW YORK — A candy sold in New York city after it was pulled from store shelves in Poland in scare over mad cow disease is safe, the U.S. Food and Drug Ad-

FDA spokesman Brad Stoner said Monday. The manufacturer of the Mamba candy said that they did have certified gelatin. U.S. food safety regulation

"There should be no problem," Stoner said.

New York City officials said Monday that one of its ingredients may have



## NEWS RELEASE

COMMERCE COMMITTEE DEMOCRATS  
Congressman John D. Dingell, Ranking Member

For Immediate Release  
February 1, 2001

Contact: Laura Sheehan  
202-225-3641

### Dingell and Waxman Question FDA's Knowledge of Dietary Supplements Containing BSE

Washington, D.C. — Congressmen John D. Dingell, Ranking Member of the Committee on Energy and Commerce, and Henry Waxman, Ranking Member of the Committee on Government Reform, in a recent letter to the Food and Drug Administration (FDA) questioned the safety and nature of dietary supplements currently sold and marketed in the United States which may contain bovine spongiform encephalopathy (BSE).

"Dietary supplements, unlike prescription drugs or vaccines, require no pre-market approval and no post-market surveillance. This makes it more difficult for the FDA to adequately oversee what products are being sold, where the products and the products' ingredients originate, and which products may contain potentially dangerous materials," the Congressmen wrote.



January 2002

## MAD COW DISEASE

Improvements in the  
Animal Feed Ban and  
Other Regulatory  
Areas Would  
Strengthen U.S.  
Prevention Efforts



**...federal actions  
do not sufficiently  
ensure that all BSE-  
infected animals or  
products are kept  
out...**



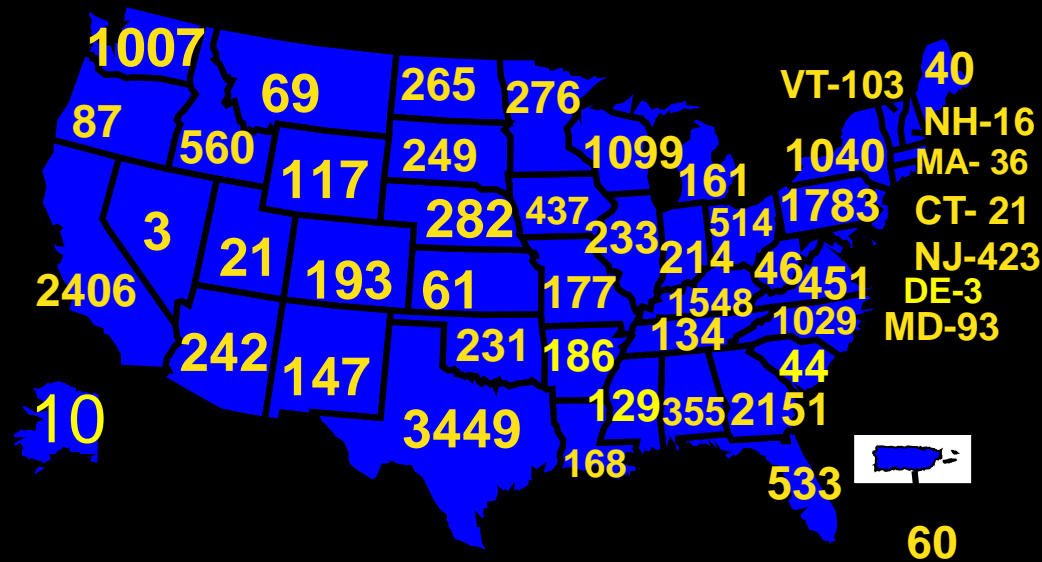
# USDA's BSE Surveillance

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- **The BSE surveillance consists of testing:**
  - **Adult cattle displaying evidence of neurological disease presenting:**
    - **On-farm (field CNS cases), veterinary hospitals, veterinary diagnostic laboratory, public health laboratories, at slaughter (CNS condemns).**
  - **“Downer cows” (nonambulatory adult cattle).**
  - **Adult cattle which die on-farm.**

# Total Bovine Brain Submissions for BSE Surveillance, United States, May 10, 1990 thru Feb 28, 2002

Total = 22,912



**No evidence of BSE detected**

Source: USDA, APHIS, NVSL



# BSE Preventive Measures

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- Since 1989, the USDA has restricted the importation of live cattle and certain cattle products from the United Kingdom.
- This restriction was later expanded to prohibit importation from all European countries and recently from Japan.
- In 1997, the Food and Drug Administration prohibited the use of most mammalian protein in the manufacture of ruminant feed .

# Harvard BSE Risk Assessment

---

- **The United States is highly resistant to introduction of BSE.**
  - **Most effective preventive measures included ban on importation of live cattle and ruminant meat and bone meal from the United Kingdom since 1989 and all of Europe since 1997, and the ruminant feed ban.**
- **BSE is extremely unlikely to become established in the United States even if it were inadvertently introduced.**

# U.S. Concerns Related to TSEs

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- Concerns related to possible introduction of BSE into the United States.
- **Concerns related to possible occurrence of vCJD and/or its possible secondary spread via blood and blood products.**
- Concerns related to CJD infection control issues and the possible zoonotic transmission of animal TSEs prevalent in the United States.

# Possible U.S. Occurrence of vCJD

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- **Primary source of exposure of the U.S. population to the BSE/vCJD agent:**
  - **During visits to BSE-endemic areas.**
  - **Consumption or use of imported products containing BSE-contaminated cattle parts.**
  - **Possible secondary person-to-person spread of the vCJD agent.**



# Blood Donor Deferral



American Red Cross

## American Red Cross Position Statement to the Transmissible Spongiform Encephalopathy (TSE) Committee

The safety of the blood supply is a top priority. The Red Cross and the American Blood Centers have taken the prudent step to ensure blood safety in the United Kingdom based on the threat of variant Creutzfeldt-Jakob disease (vCJD).

The current deferral is for people who have lived in the United Kingdom (England, Northern Ireland, Scotland) more than six months between 1980 and 1996. The deferral also includes France as well as all of Western Europe. The American Blood Centers and the American Red Cross have urged the Spongiform Encephalopathy (BSE) in the United Kingdom. The TSE committee should recommend a deferral of less than six months in the U.K. We believe that the current exposure period between 1980 to the present is too long.

July 6, 2001

## U.S. Urges Use of Blood From Military

By RAYMOND HERNANDEZ

**W**ASHINGTON, July 5 — As it moves to restrict blood donations from people who might have been exposed to mad cow disease in Europe, the U.S. military is urging a change in the way the nation handles its blood supply.

### RELATED ARTICLES

[New York Health Commissioner Favors Blood Restrictions \(June 28, 2001\)](#)

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[Blood Supplies Critically Short in N.Y. Region \(June 25, 2001\)](#)



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July 3, 2001

## FDA Advisory Committee tightens restrictions on blood donation

Red Cross, America's Blood Centers fear tighter restrictions will continue to deplete supply

A Food and Drug Administration (FDA) advisory committee voted June 28 to further tighten the criteria for deferring blood donors who have lived in Europe. The vote reflected concerns that variant Creutzfeldt-Jakob disease (vCJD), the human form of Bovine Spongiform Encephalopathy (BSE) or "mad cow" disease, might be spread through blood transfusions.

Although the tighter restrictions are intended to protect America's blood supply, donation organizations such as the Red Cross and America's Blood Centers fear excessive restrictions would further exacerbate the shortage of blood in the U.S. During the meeting, both organizations put forward their own "compromise" restrictions, which they said would provide the best balance between supply and protection.

Current FDA guidelines, implemented by all U.S. blood centers in 1999, prevent blood donations from people who have spent more than six months in Great Britain, considered the hub of BSE activity, between 1980 and 1996. vCJD is believed to be transmitted to humans from infected cattle. (see Washington Fax 4/24/2001b)

restrictions in the New York City area.

states that the shortage of blood from its metropolitan region, where medical officials will worsen the existing blood shortage and a variety of surgeries.

proposed by the Food and Drug Administration



# Blood Donor Deferral Policy

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- In 1999, FDA instituted a policy to defer blood donors who spent a cumulative  $\geq 6$  months period in the United Kingdom during 1980-1996.
- Because of the emergence of BSE and vCJD in other European countries, FDA recently expanded the blood donor deferral policy.
  - UK policy tightened from cumulative  $\geq 6$  months to  $\geq 3$  months.
  - Excludes donors who spent  $\geq 5$  years in other European countries, and persons who lived on US military bases in Europe.

# Blood Donor Deferral Policy

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- The donor deferral policy is intended to minimize the risk of vCJD transmission, not totally eliminate the risk.
- The length of time spent in Europe was selected to maximize the benefit of the deferral policy with minimum adverse impact on blood availability.
- The donor deferral criteria were estimated to result in a 90% reduction in total person-days of risk-weighted exposure to the vCJD agent.

# Blood Donor Deferral Policy

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- **The risk of vCJD transmission via blood and blood products is considered theoretical because:**
  - **No transmission of the vCJD agent by human blood or plasma has been reported.**
  - **Study of recipients of blood products from vCJD donors has not demonstrated infectivity.**

# Why the Concern About Bloodborne Transmission?

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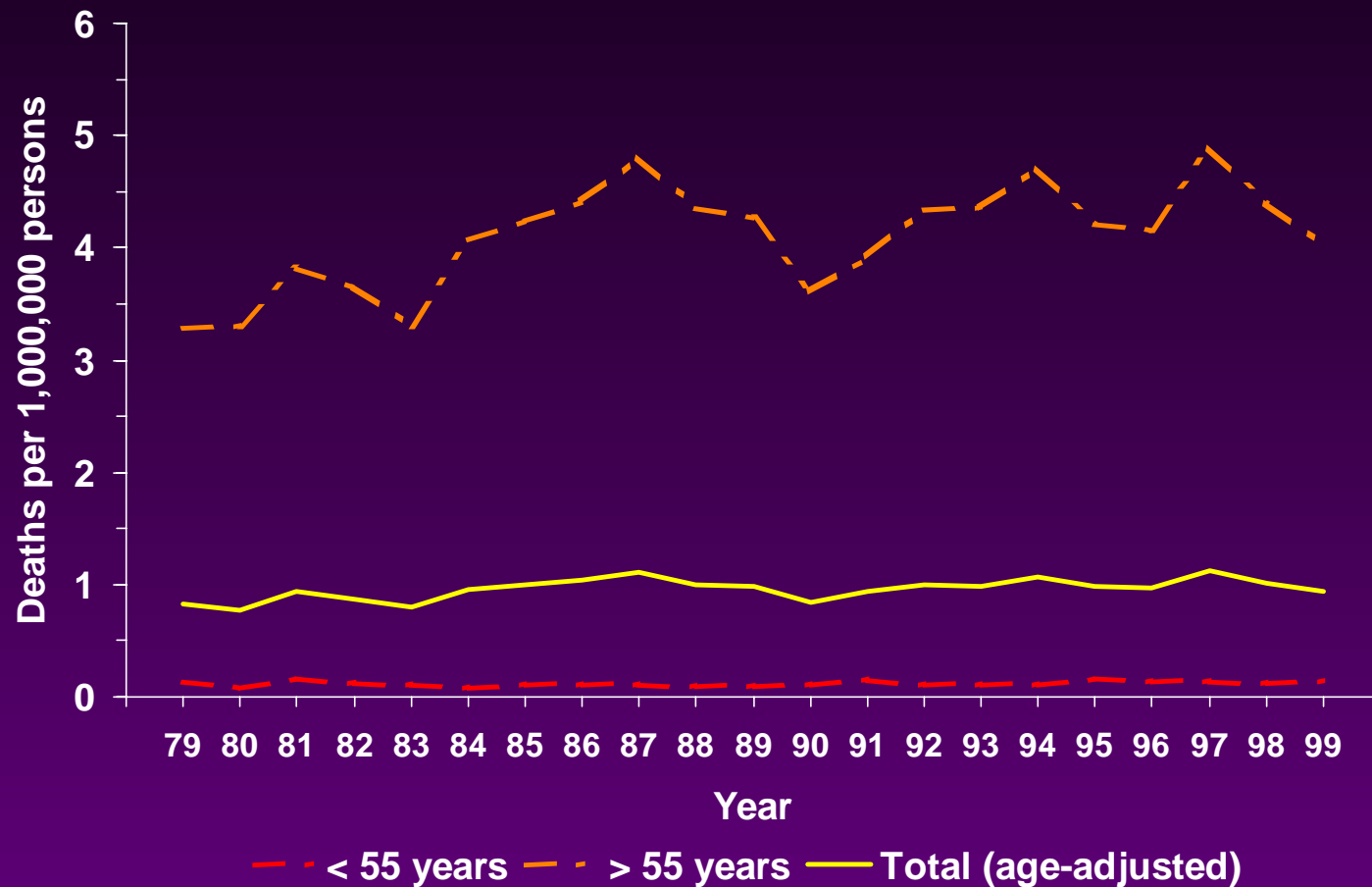
- The vCJD agent is readily detectable in lymphoreticular tissues.
- Presumed route of BSE transmission through ingestion implies possible blood phase – prionemia.
- Transmission of the BSE agent via transfusion during the incubation period in an experimental sheep model has been reported.
- Variant CJD is a new fatal disease with very long incubation period.

# CJD/vCJD Surveillance

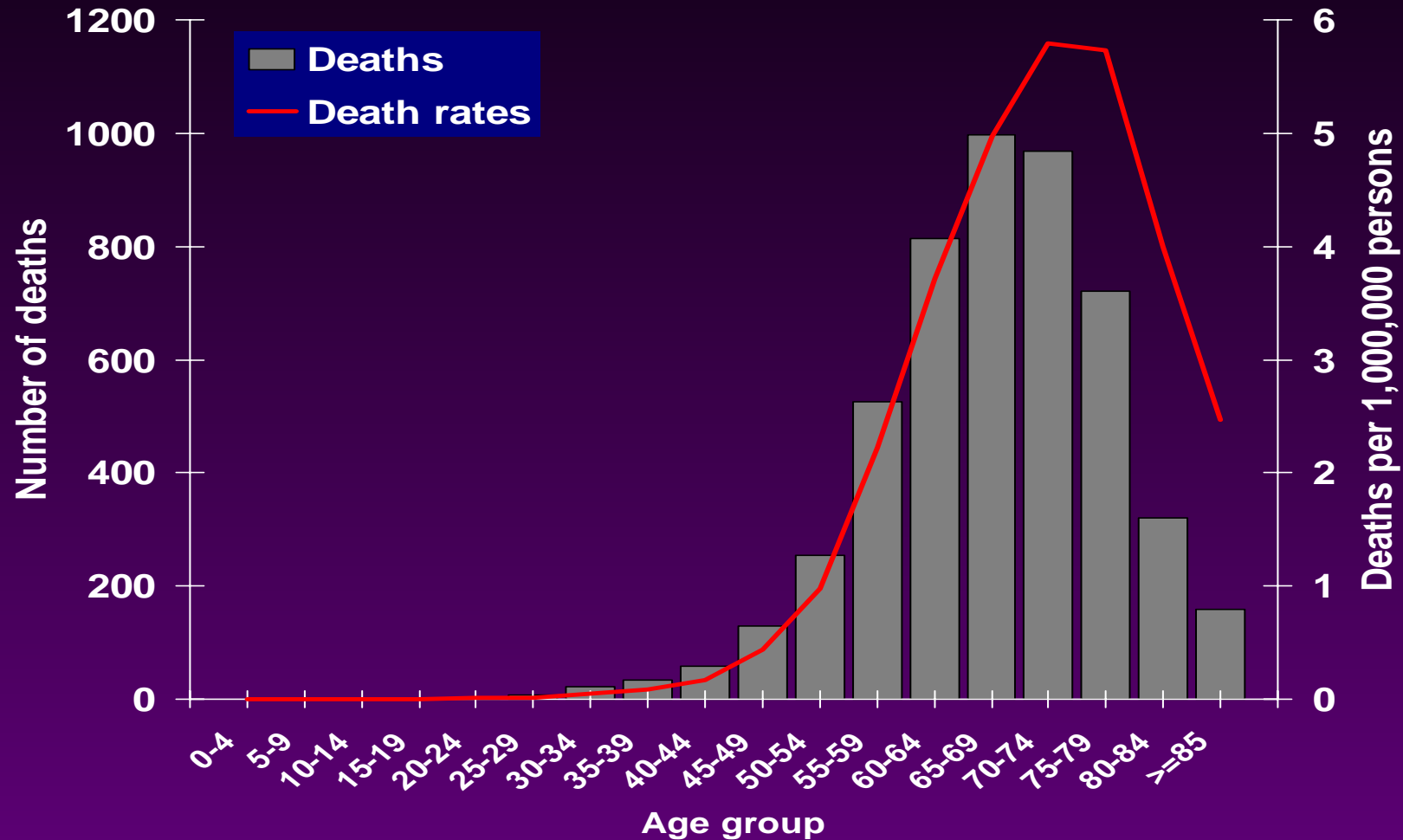
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- In 1996, CDC enhanced surveillance to monitor CJD and the possible occurrence of vCJD.
  - Periodic review of the national multiple cause-of-death data.
  - Follow up investigation of CJD decedents <55 years of age in collaboration with state and local health departments.
  - Supporting the establishment of the National Prion Disease Pathology Surveillance Center in collaboration with the American Association of Neuropathologists.

# Age-Adjusted and Age-Specific CJD Death Rates, United States, 1979-1999

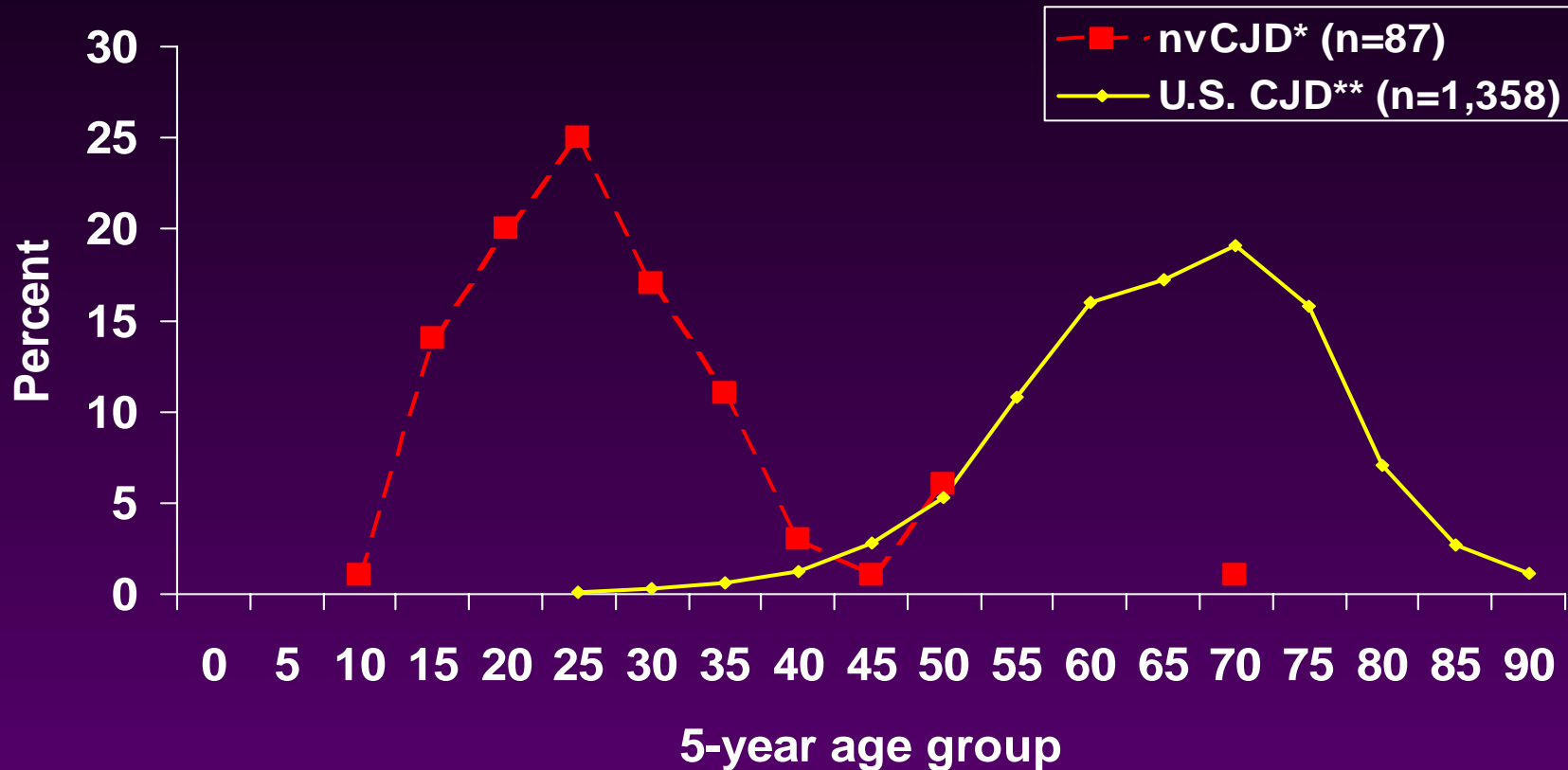


# CJD Deaths and Death Rates by Age-Group, United States, 1979-1999





# Percent Distribution of U.K. vCJD and U.S. Sporadic CJD Cases by Age Group at Death, 1995-2001



\*Data as of May 1, 2001

\*\*Non-iatrogenic cases, 1995-1999

# Cases Received and Diagnosed by the National Prion Disease Pathology Surveillance Center\*

Year	Referrals	Prion disease				Prion disease (total)
		Sporadic	Familial	Iatrogenic	vCJD	
1997	104	54	6	0	0	60
1998	94	44	6	1	0	51
1999	114	65	9	0	0	74
2000	169	97	12	2	0	111
2001	244	136	16	0	0	152

\*Information available at [www.cjdsurveillance.com](http://www.cjdsurveillance.com)



# U.S. Concerns Related to TSEs

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# Infection Control Concerns

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**Patients possibly given brain disease**  
July 11, 2001 Posted: 12:31 PM EDT (1631 GMT)

**CHICAGO, Illinois (AP) –** Fourteen patients at two hospitals may have been exposed to a rare and deadly brain disease through tainted surgical instruments, according to an alert issued Tuesday to hospitals nationwide.

The patients at both hospitals, Exempla St. Joseph in Denver, Colorado, and Tulane University Hospital and Clinic in New Orleans, Louisiana, underwent surgery in which instruments were used that had been previously used on people later diagnosed with

Creutzfeldt-Jakob disease. Neither case involved the form of the ailment that may be related to mad cow disease, according to the Joint Commission on Accreditation of Healthcare Organizations. The nonprofit group sent the alert to the nearly 5,000 hospitals nationwide that it monitors and accredits. Because normal sterilization techniques don't kill the organism that causes the disease, the commission said all surgical instruments used in neurosurgery when a diagnosis

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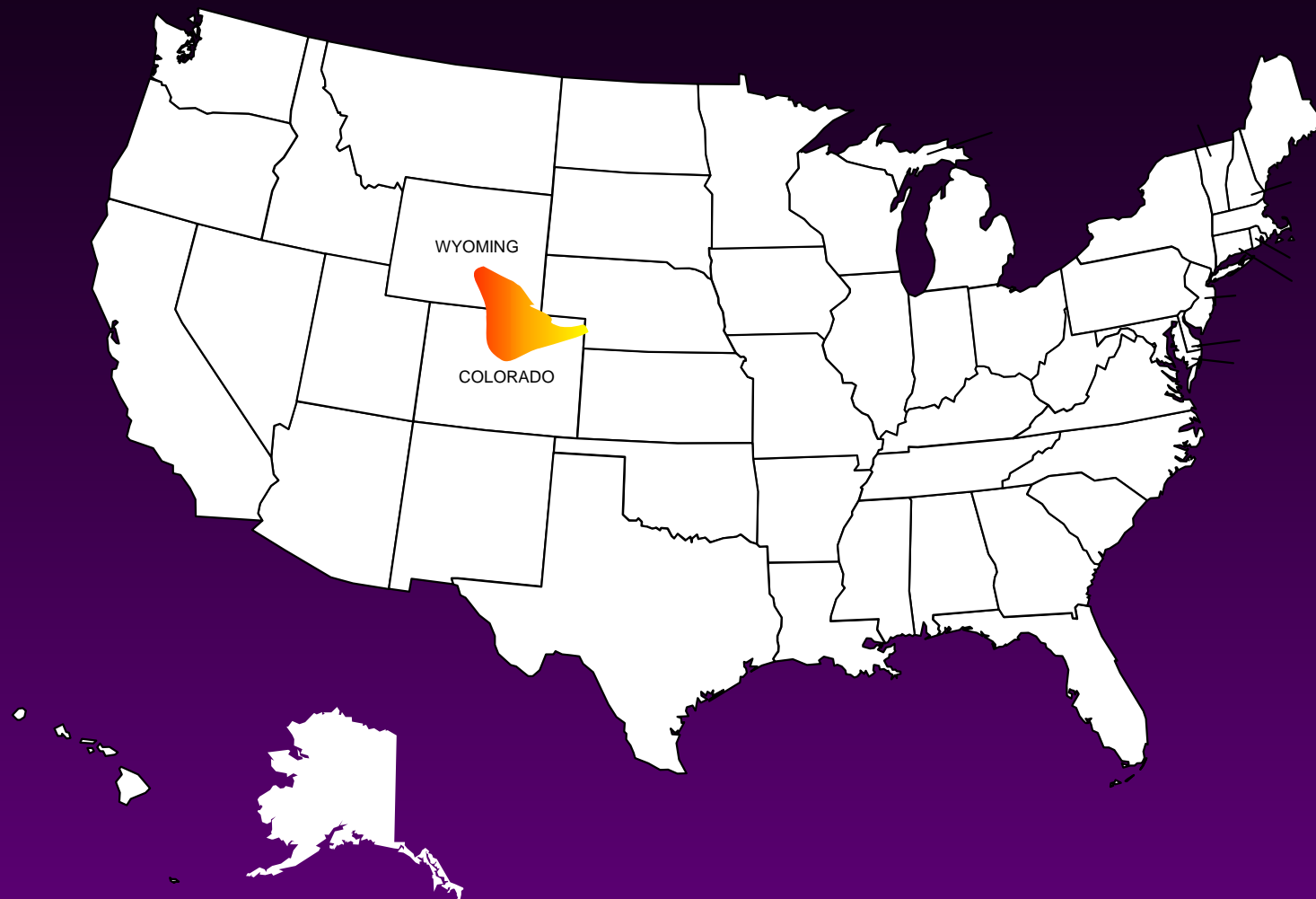




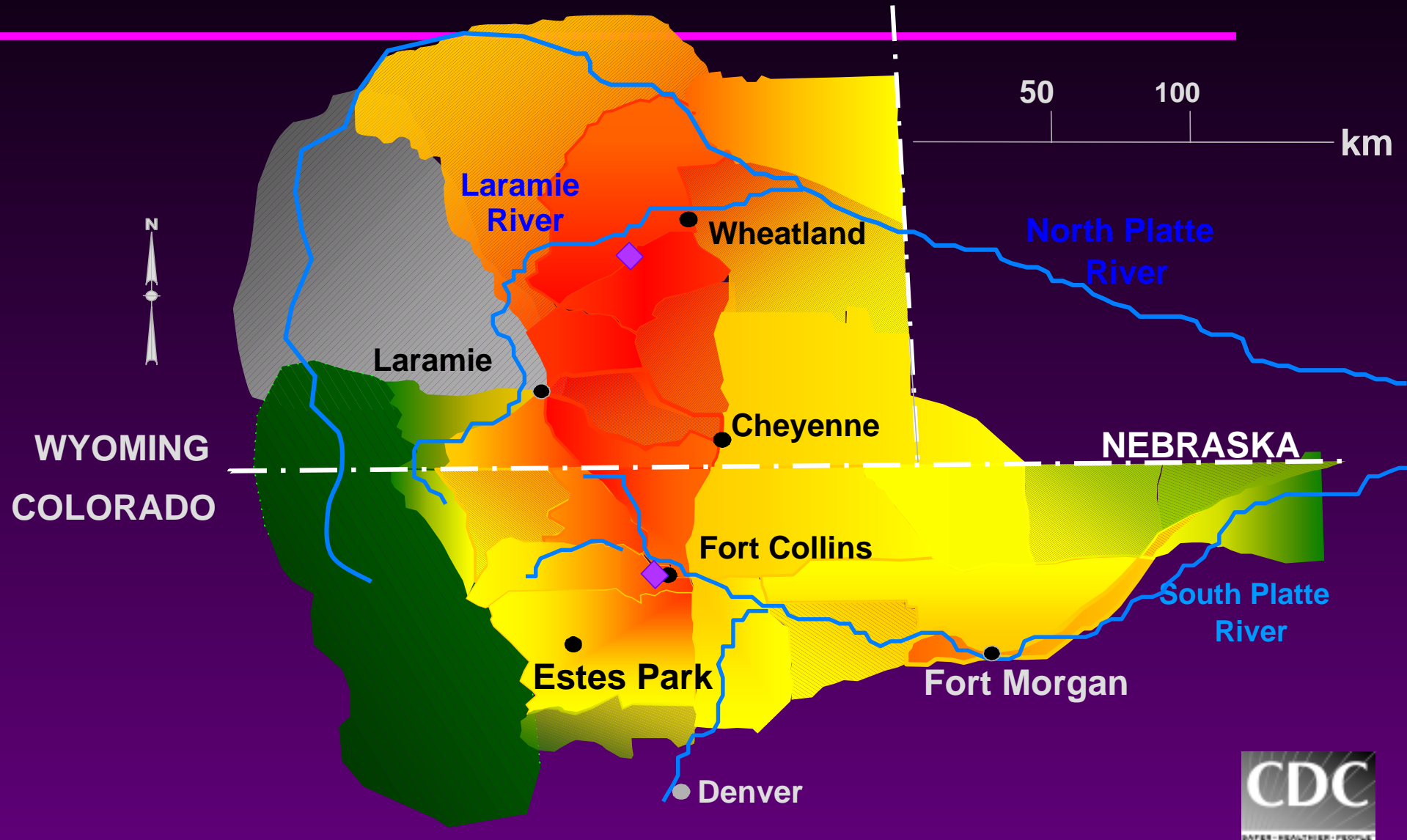
**CDC**

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# Chronic Wasting Disease Endemic Areas, United States

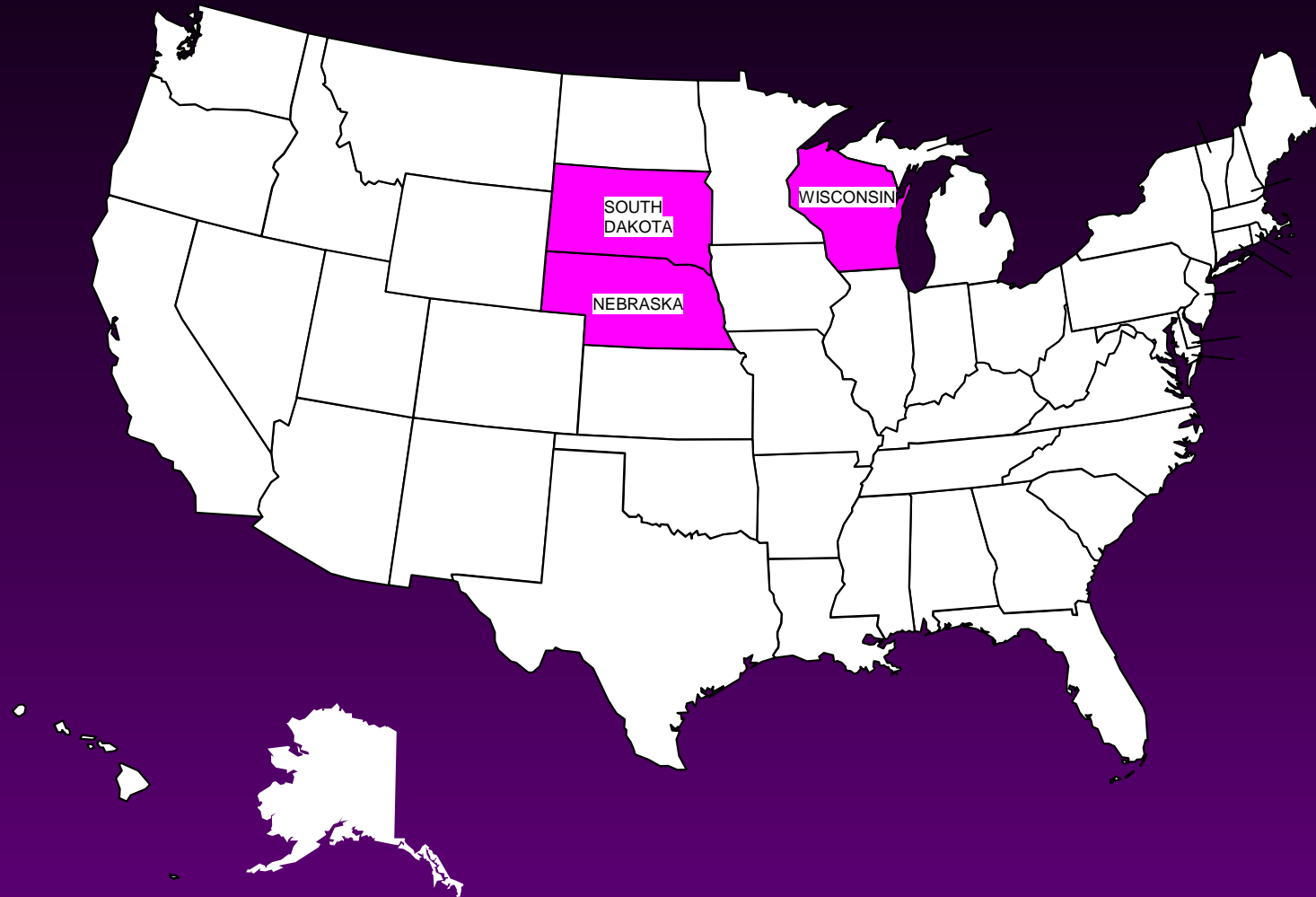


# Chronic Wasting Disease Endemic Areas, United States



# States Where CWD has Recently been Identified Among Free-Ranging Deer or Elk

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# Clinical Characteristics of the 3 Unusually Young CJD Patients with Venison Consumption

	Case 1	Case 2	Case 3
<b>Year of death</b>	1997	1999	2000
<b>Age at death</b>	28	30	28
<b>Sex</b>	Female	Male	Male
<b>Presentation</b>	Abnormal mental status, unsteady gait	Cognitive difficulties	Memory loss, behavioral change, confusion
<b>Illness duration</b>	4 mos	10 mos	15 mos
<b>EEG</b>	Not classic	No abnormality	Not classic

# Comparison of Key Evidence Supporting a Causal Link Between BSE and vCJD to that of CWD and 3 Unusually Young CJD Cases in the United States.

---

	<b>BSE/vCJD</b>	<b>CWD/CJD in the 3 Patients</b>
<b>Increasing incidence of young cases</b>	<b>Definite</b>	<b>Not definite</b>
<b>Distinctive neuropathology</b>	<b>Yes</b>	<b>No</b>
<b>Phenotypic homogeneity</b>	<b>Yes</b>	<b>No</b>
<b>PrP-res different from classic forms</b>	<b>Yes</b>	<b>No</b>
<b>Definite food consumption from endemic/epidemic areas</b>	<b>Yes</b>	<b>No</b>
<b>Polymorphism at codon 129</b>	<b>Met/Met</b>	<b>Heterogeneous</b>

---

# Case Investigation Summary

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- The occurrence of three unusually young CJD patients suggested a possible relationship with CWD.
- Our investigation found no conclusive evidence for a causal link between CWD and CJD in the patients.
- Continued surveillance remains critical to monitor the possible transmission of CWD to humans.

# Additional Case Investigations

---

- Investigated a 25-year-old patient who consumed venison originating from Southeastern Wyoming.
  - He was later shown to have GSS 102 mutation with valine at codon 129 in the mutant allele of the prion protein gene.
- Investigated two patients (26 and 28 years of age) who lived in adjacent counties and had illness onset within several months of each other.
  - Although venison consumption was reported for the 28-year-old patient at ~1.5 years of age, this history was questionable; the immunohistochemical analysis was consistent with GSS.
  - No venison consumption history was reported for the 26-year-old patient.

# Summary

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**Continued surveillance is critical to monitor the possible occurrence of BSE and vCJD in the United States as well as monitor the risk, if any, of CWD transmission to humans.**