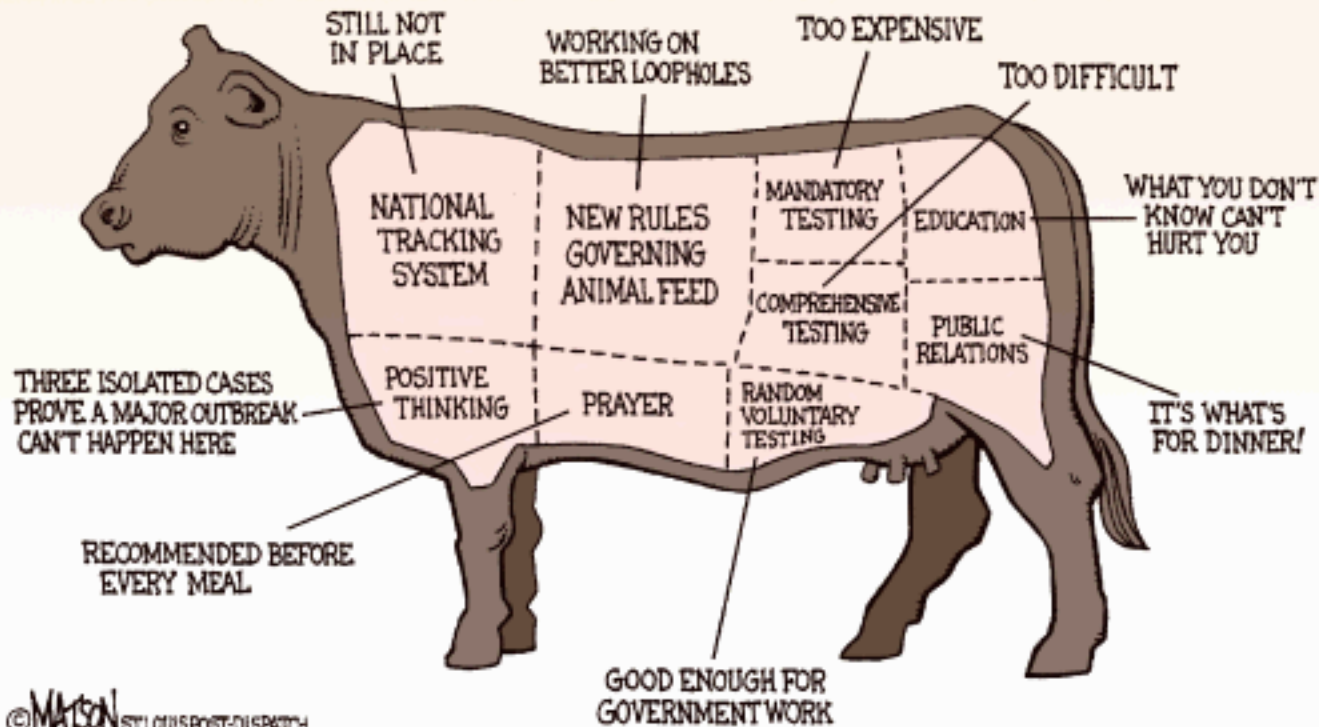


# Lecture 19 – BSE and vCJD

## MAD COW DISEASE

*And What The U.S.D.A. Is Doing About It*





## Stanley Prusiner, UCSF

**Science** The World's Leading Journal of Original Scientific Research, Global News, and Commentary.

Science, Vol 216, Issue 4542, 136-144  
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### ARTICLES

#### **Novel proteinaceous infectious particles cause scrapie**

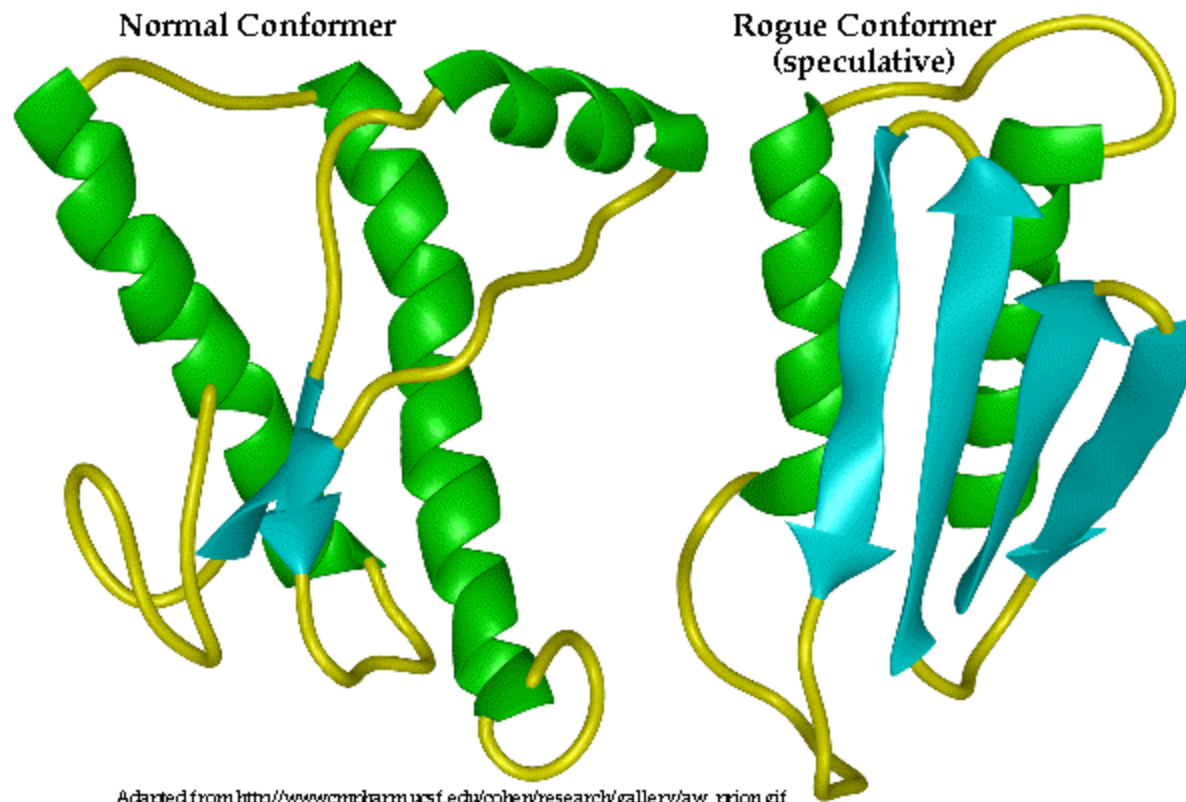
SB Prusiner

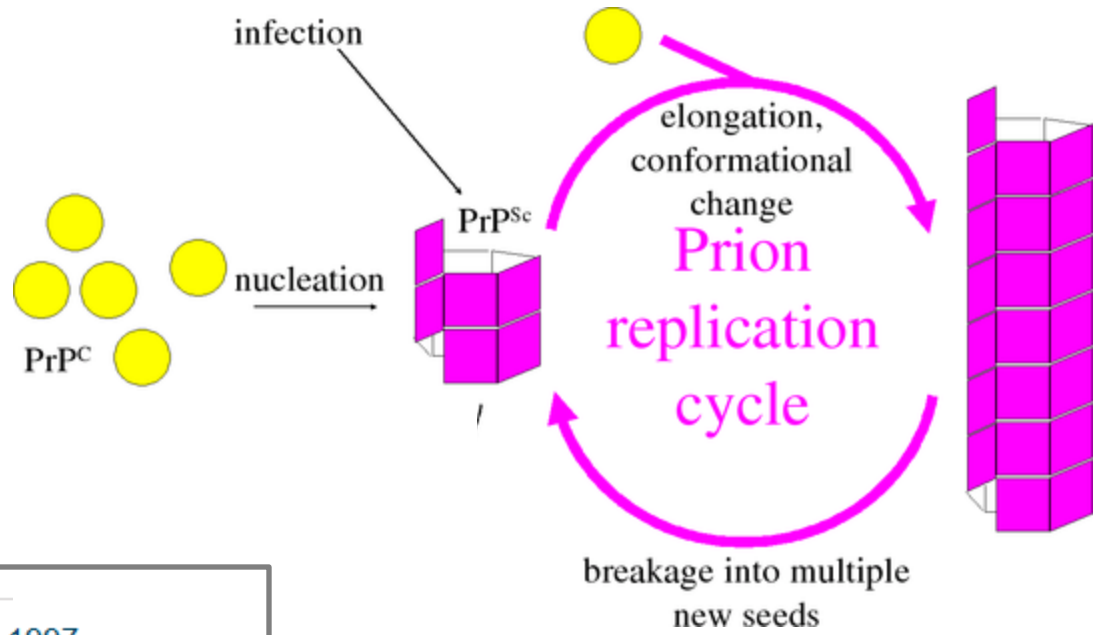
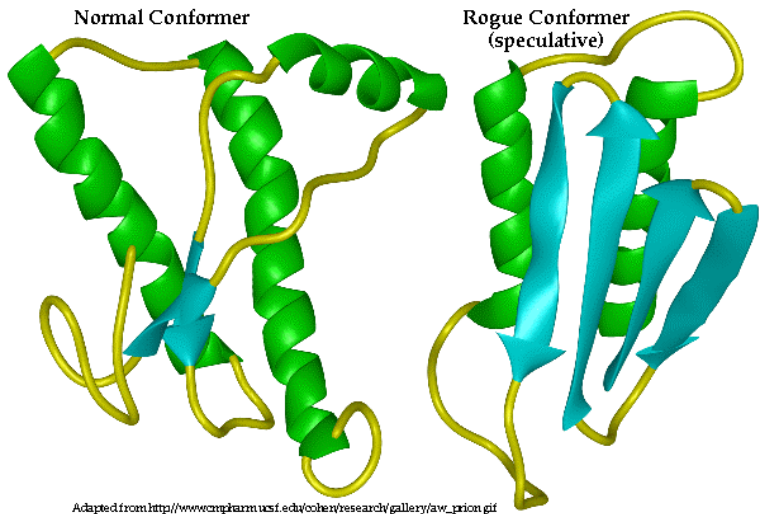
After infection and a prolonged incubation period, the scrapie agent causes a degenerative disease of the central nervous system in sheep and goats. Six lines of evidence including sensitivity to proteases demonstrate that this agent contains a protein that is required for infectivity. Although the scrapie agent is irreversibly inactivated by alkali, five procedures with more specificity for modifying nucleic acids failed to cause inactivation. The agent shows heterogeneity with respect to size, apparently a result of its hydrophobicity; the smallest form may have a molecular weight of 50,000 or less. Because the novel properties of the scrapie agent distinguish it from viruses, plasmids, and viroids, a new term "prion" is proposed to denote a small proteinaceous infectious particle which is resistant to inactivation by most procedures that modify nucleic acids. Knowledge of the scrapie agent structure may have significance for understanding the causes of several degenerative diseases.

Prion – **Protein Infection**

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# Prion – Protein Infection

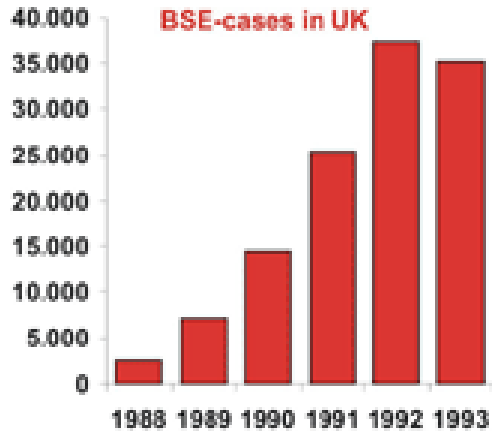




The Nobel Prize in Physiology or Medicine 1997  
Stanley B. Prusiner

The Nobel Prize in Physiology or Medicine 1997

The Nobel Prize in Physiology or Medicine 1997 was awarded to Stanley B. Prusiner "for his discovery of Prions - a new biological principle of infection".



For each diagnosed case, six animals entered the human food chain.

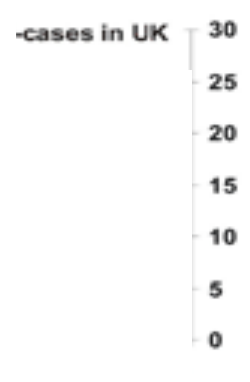
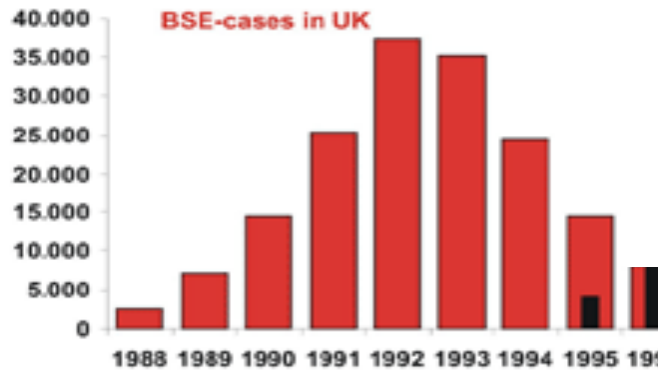


A sign in a British McDonald's restaurant

*'The safety of beef is not in question.'*

*'There is no scientific evidence that BSE can be transmitted to humans.'*

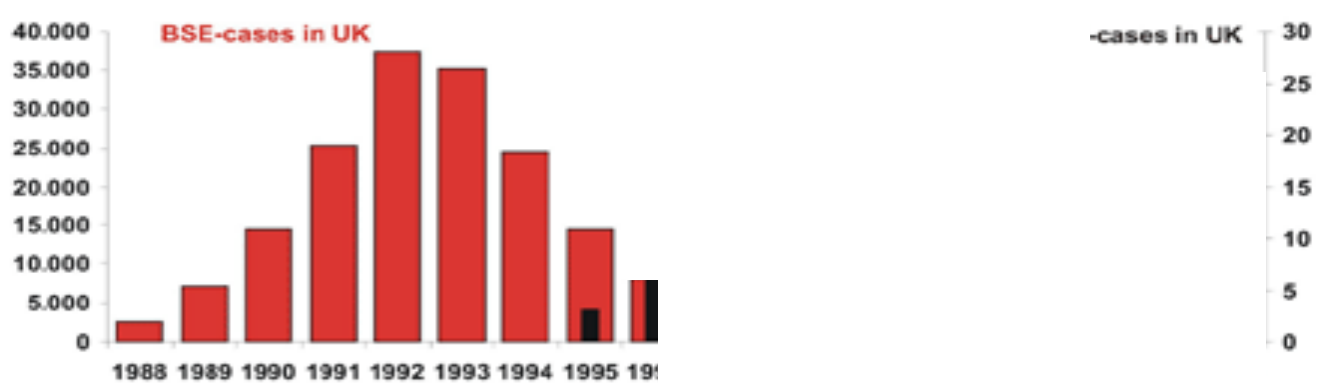
Prime Minister John Major in 1995





*‘There remains no scientific proof that BSE can be transmitted to man by beef, but the committee have concluded that the most likely explanation at present is that these cases are linked to exposure to BSE before the introduction of the specified bovine offal ban in 1989.’*

Health Minister Stephen Dorrell, March 20 1996



## List of some vCJD mortality estimates

1990	'A generation'	Dr Richard Lacey
1996	'Hundreds'	UK Government
1996	'zero to millions'	Royal Statistical Society
1997	'75 to 80,000'	Nature paper
2000	'up to 250,000'	Phillips enquiry

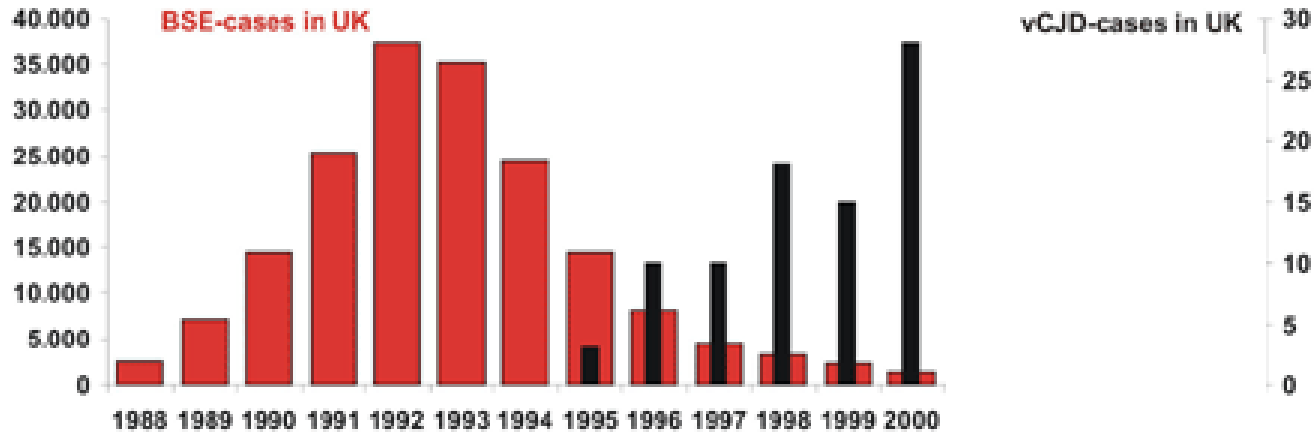
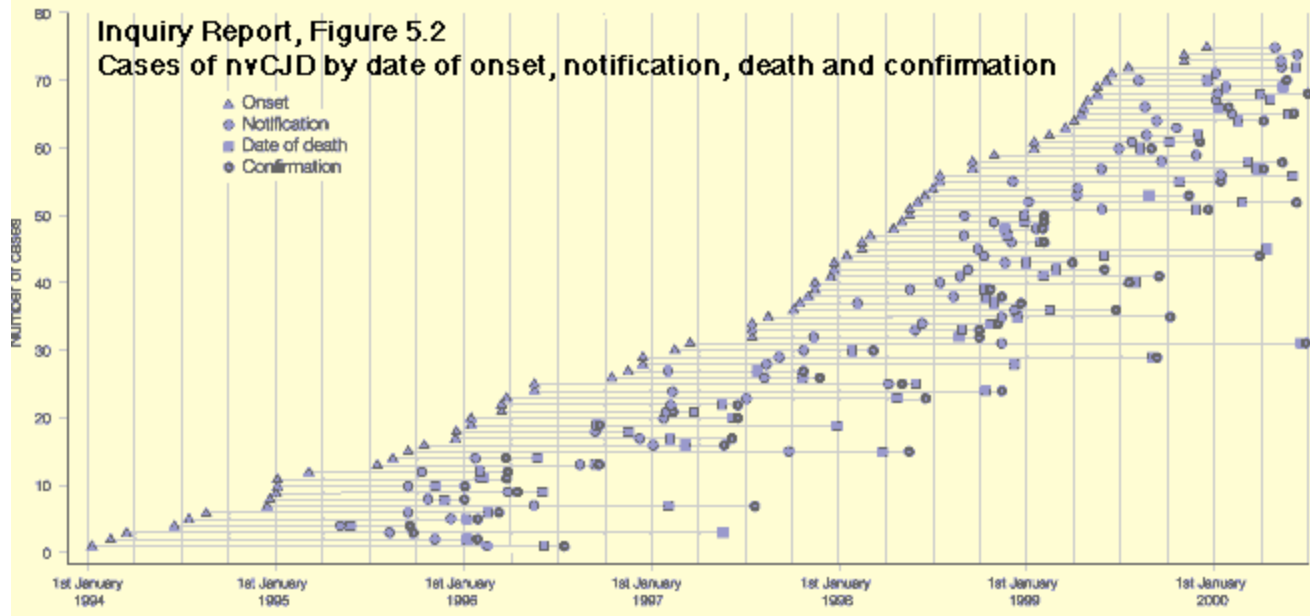
*It is 20 March 2016. Britain's euthanasia clinics churn on overtime, struggling to help 500 people a week to a dignified death before brain disease robs them of reason and self control.*

*This nation, whose leaders spent a decade in denial, is now quarantined, the world having long since shunned contact with a population in which half a million people a year succumb to Creutzfeldt-Jakob disease. The channel tunnel is blocked with 5 miles of French concrete.*

The Observer newspaper predicts a grim future in 1996.

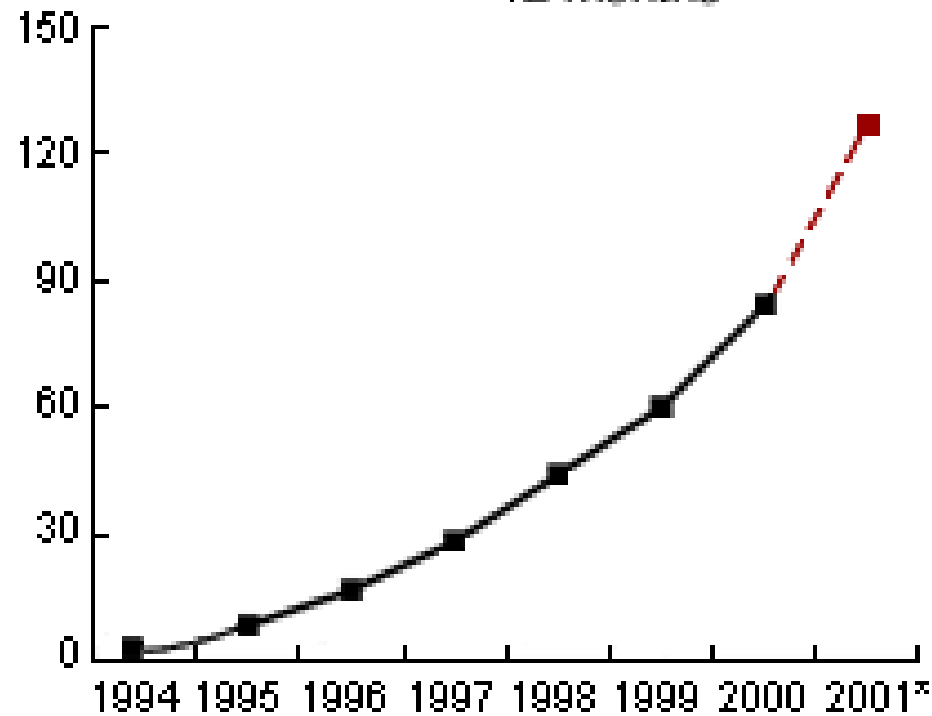
### Inquiry Report, Figure 5.2

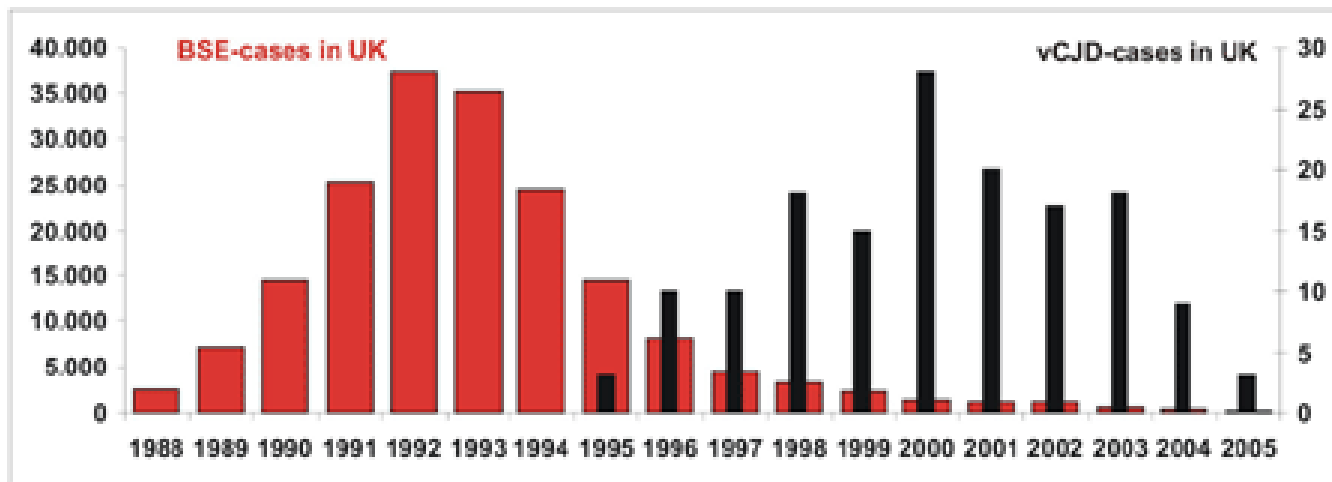
### Cases of nvCJD by date of onset, notification, death and confirmation



# Britain's nvCJD toll accelerating

predictions for next 12 months

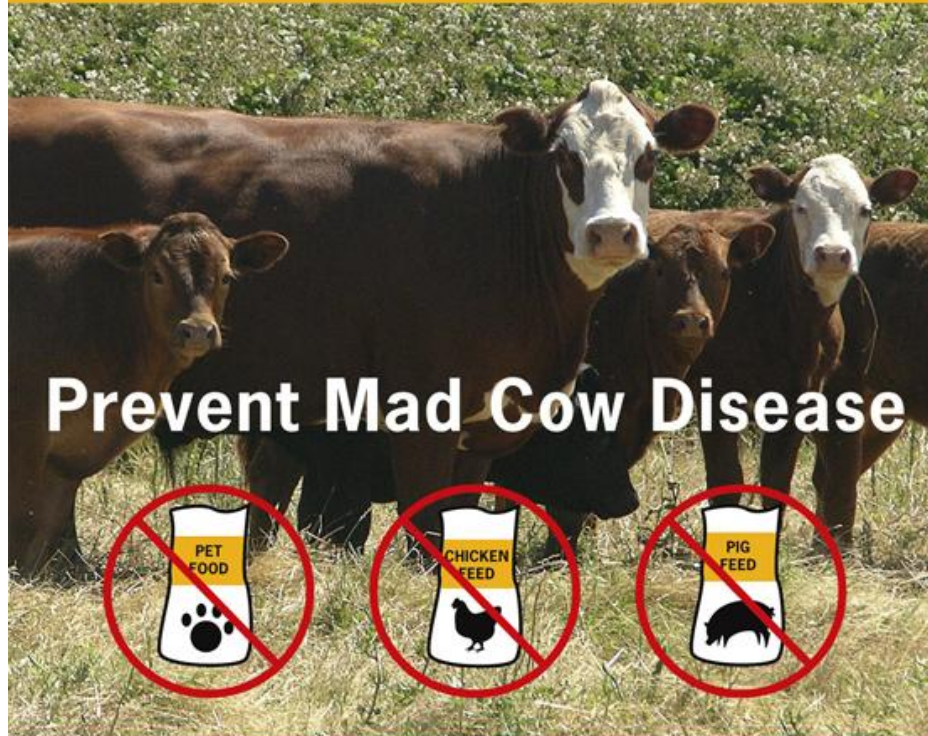






# Help Keep California Cows Healthy

**CAUTION:** FEED LABELED,  
"DO NOT FEED TO CATTLE OR  
OTHER RUMINANTS," CONTAINS  
PROHIBITED ANIMAL PROTEIN\*,  
WHICH CAN CAUSE MAD COW  
DISEASE IF FED TO CATTLE.



## PROHIBITED CATTLE FEED (RUMINANT FEEDING BAN)

- Pet food is not labeled with the above caution statement; however, it may contain prohibited animal protein\* and should not be fed to cattle or other ruminants.
- \*Prohibited animal protein is any protein-containing portion of mammalian animals, excluding: blood products; gelatin; inspected meat products that have been cooked and further heat processed for feed (such as plate waste); milk products; and any product that consists entirely of porcine (pork) or equine (horse) protein.

For more information call the California Department of Food and Agriculture's Safe Animal Feed and Education Program (SAFE) at 916-445-0444 or visit us on the web at [www.cdffa.ca.gov/is/safe](http://www.cdffa.ca.gov/is/safe).



25 thousand cattle .....

2004: 15.5 thousand

20 .....

**Number of cattle  
tested for mad  
cow disease,  
by year**  
May 1990 - April 2004

15 .....

10 .....

5 .....

1990: 40 cows

0

'90 '92 '94 '96 '98 '00 '02 '04\*

.....

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# U.S.

## First apparent U.S. case of mad cow disease discovered

### Japan, other nations ban import of U.S. beef

Wednesday, December 24, 2003 Posted: 0722 GMT ( 3:22 PM HKT)

**WASHINGTON (CNN) -- The first apparent case of mad cow disease in the United States has been discovered, the U.S. Department of Agriculture said Tuesday.**

Two tests have been carried out on meat from the cow, enabling Agriculture Secretary Ann Veneman to call the case a "presumptive positive." A sample is being flown to England for a third test to confirm the case.

Bovine spongiform encephalopathy, or BSE, is linked to a similar form of the incurable and fatal brain-wasting disease in humans, called variant Creutzfeldt-Jakob Disease, or vCJD. There have been a small number of cases of vCJD reported worldwide, primarily in the United Kingdom, in people who ate BSE-contaminated meat.

Within hours of the announcement, an official with Japan's agriculture ministry told CNN that his country would ban imports of U.S. beef. South Korea, Taiwan, Malaysia and Singapore all followed suit Wednesday.



Mad cow disease causes severe nervous system deterioration and has been linked to a similar disease in humans.

Mad cow disease causes severe nervous system deterioration and has been linked to a similar disease in humans.

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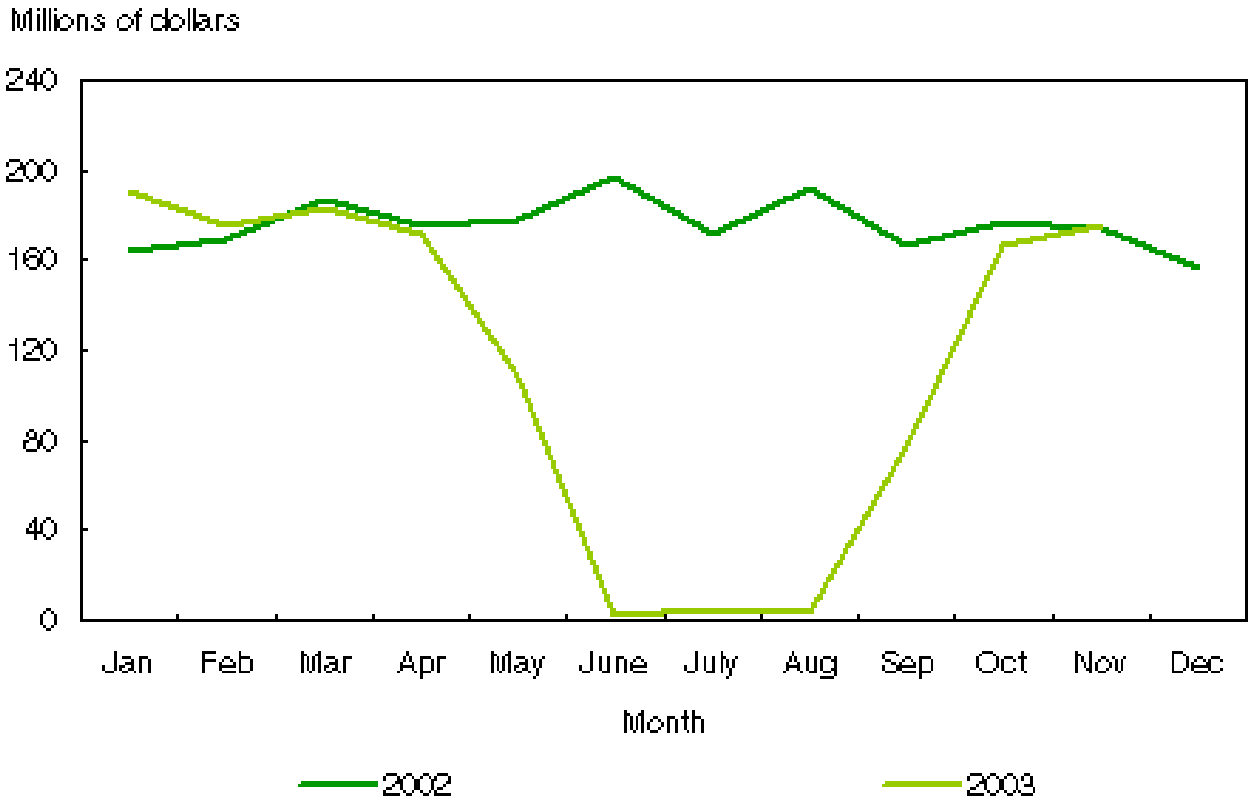
CNN's Christy Feig reports on the first apparent case of mad cow disease in the United States.

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# Canadian beef exports after the discovery of a single BSE infected cow in May 2003



Source: International Trade Division, Statistics Canada.





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## Court: US can block mad cow testing

Posted 8/29/2008 1:42 PM | [Comment](#) | [Recommend](#)

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By **Matt Apuzzo**, Associated Press Writer

WASHINGTON — The Bush administration can prohibit meat packers from testing their animals for mad cow disease, a federal appeals court said Friday.

The dispute pits the Agriculture Department, which tests about 1 percent of cows for the potentially deadly disease, against a Kansas meat packer that wants to test all its animals.

Larger meat packers opposed such testing. If Creekstone Farms Premium Beef began advertising that its cows have all been tested, other companies fear they too will have to conduct the expensive tests.

The Bush administration says the low level of testing reflects the rareness of the disease. Mad cow disease has been linked to more than 150 human deaths worldwide, mostly in Great Britain. Only three cases have been reported in the U.S., all involving cows, not humans.

A federal judge ruled last year that Creekstone must be allowed to conduct the test because the Agriculture Department can only regulate disease "treatment." Since there is no cure for mad cow disease and the test is performed on dead animals, the judge ruled, the test is not a treatment.

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The U.S. Court of Appeals for the District of Columbia Circuit overturned that ruling, saying diagnosis can be considered part of treatment.

"And we owe USDA a considerable degree of deference in its interpretation of the term," Judge Karen LeCraft Henderson wrote.

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Tuesday, January 20, 1998 Published at 08:22 GMT

## World

# Oprah Winfrey faces mad cow libel suit



Oprah Winfrey is in Amarillo, Texas \$12m libel trial after she claimed on never eat another burger because of

The area around Amarillo, Texas wh place produces 25 percent of Americ and the city's biggest private employ slaughterhouse.

