Transmissible Spongiform Encephalopathies

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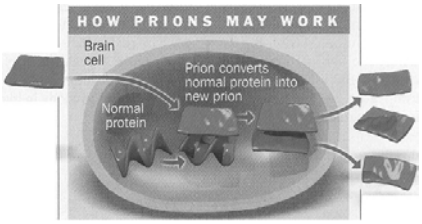
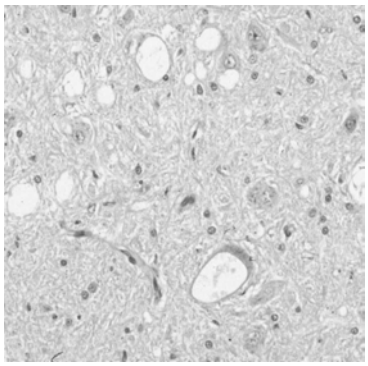
Transmissible Spongiform Encephalopathies

- Scrapie
- Kuru, and other human diseases
- Transmissible mink encephalopathy
- BSE
- CWD

TSE Pathogenesis

- all are caused by "prions"
  proteinaceous infectious particle





Scrapie

- First described in 1772
- Experimental transmission proven by accident, 1937
- Spread from infected ewe to lambs, through placental fluids
- 2-5 year incubation
- Progressive neurologic disease
- Diagnosed in U.S., 1947

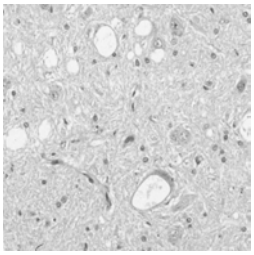
Human Spongiform Encephalopathies

- Creutzfeld-Jacob Disease (CJD)
- Kuru
- Gerstmann-Straussler-Scheinker Syndrome
- Fatal Familial Insomnia
- vCJD

Transmissible Mink Encephalopathy

- Ranched mink, Europe and the U.S.
- Behavioral changes, aggression, ataxia
- Exposure to infectious agent in feed?

Bovine Spongiform Encephalopathy
"mad cow disease"



- First noted in cattle in U.K. 1987
- Source was "infected" meat and bone meal
- Caused by a prion agent

BSE - History

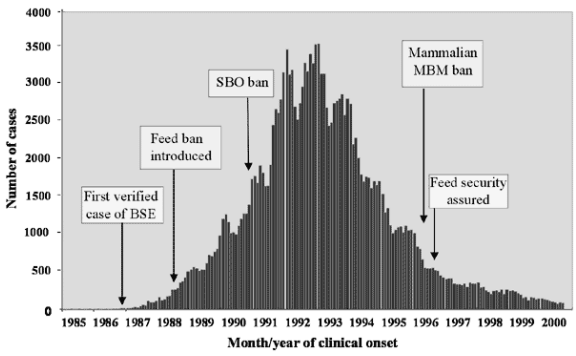
- First clinical signs, 1985
- First reported, 1987
- Reportable disease, 1988

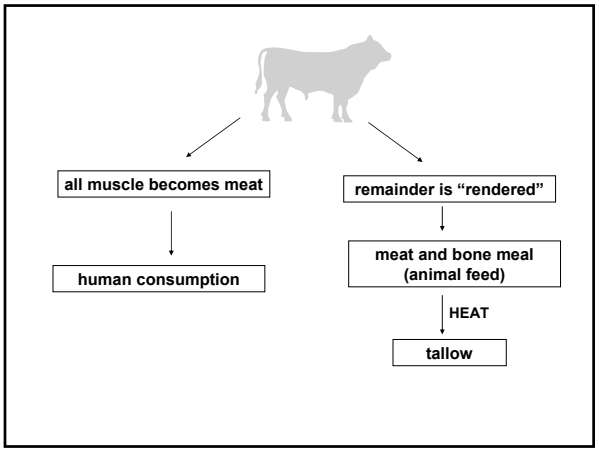
BSE – Clinical Signs

- Average incubation period is 5 years
- Unsteady gait
- Hyperesthesia
- Drop in milk yield

BSE in U.K.

- 180,000 cattle

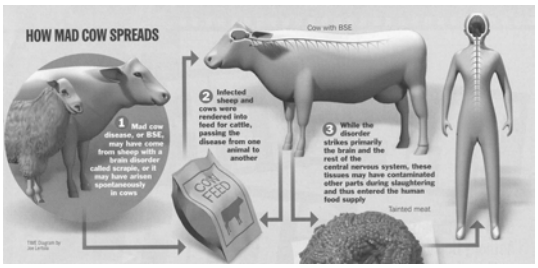




BSE – A Zoonosis?

- March 20, 1996
  cluster of unusual CJD cases

Spongiform Encephalopathy Advisory Committee



Which parts are "infectious"?

- Naturally infected cattle – brain, spinal cord, retina
- Experimentally infected cattle – all of above PLUS distal ileum, bone marrow, dorsal root ganglia

	vCJD	CJD
age	young	old
disease course	prolonged	rapid
clinical disease	behavioral changes	dementia
PrP plaques	frequent	infrequent

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Rapid Communication

Prevalence of lymphoreticular prion protein accumulation in UK tissue samples

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Abstract

This study aims to provide an estimate of the number of individuals in the UK who may be incubating variant Creutzfeldt-Jakob disease and at risk of causing iatrogenic spread of the disease. Lymphoreticular accumulation of prion protein is a consistent feature of variant Creutzfeldt-Jakob at autopsy and has also been demonstrated in the pre-clinical phase. Immunohistochemical accumulation of prion protein in the lymphoreticular system remains the only technique that has been shown to predict neurological disease reliably in animal prion disorders. In this study, immunohistochemistry was used to demonstrate

16,700 appendectomy and tonsillectomy samples from 20-29 year olds

3 were positive, therefore 237 cases per million

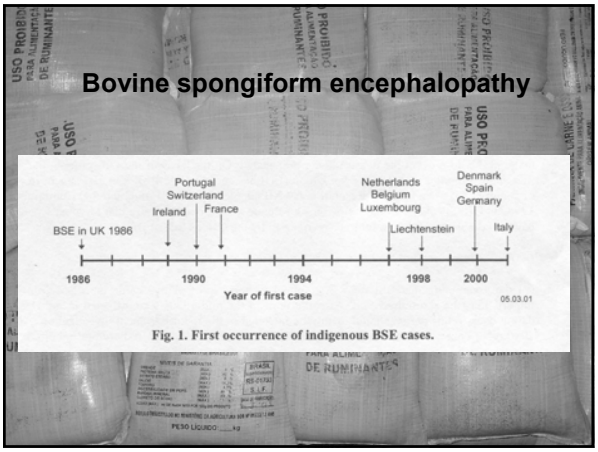


Fig. 1. First occurrence of indigenous BSE cases.

Germany rues 'complacency' over BSE testing strategy — *Nature*, November 30, 2000

POLICY AND PEOPLE

Germany's Health and Agriculture ministers resign over handling of BSE

Lancet, January 20, 2001

Failure to recognize, failure to act…..

CLE

Career Limiting Event

Will BSE continue to spread?

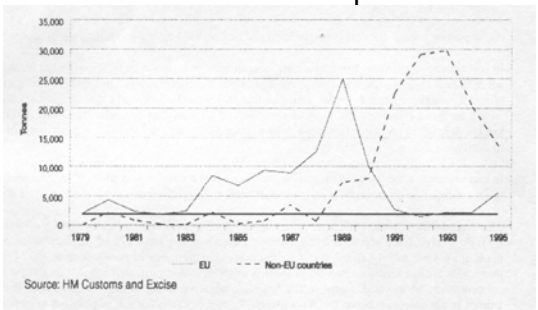
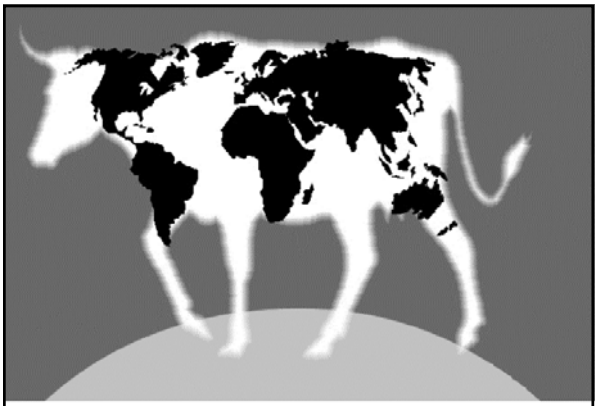
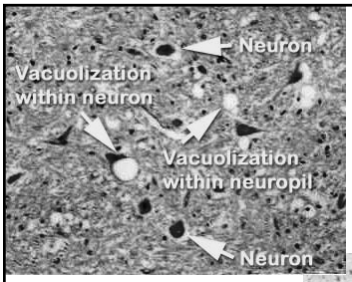


Fig. 2. UK exports of flours, meals and pellets of meat or meat offal, unfit for human consumption (greaves), 1979–1995.

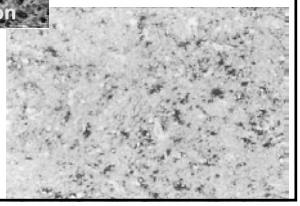


There is no live animal test



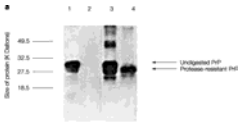
←histopathology

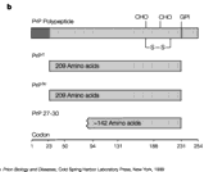
immunohistochemistry →

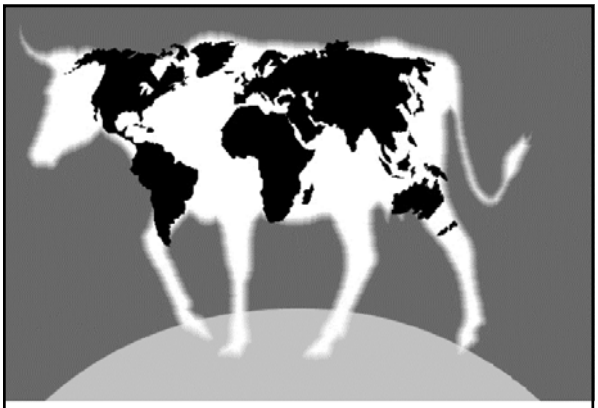


Rapid Tests

- Western blot
- ELISA







U.S. bans Canadian beef after mad cow discovery

Case in Alberta called isolated

By Elizabeth Weise
USA TODAY

The United States banned all imports of cattle, beef and beef products from Canada Tuesday, after a cow in Alberta tested positive for mad cow disease.

Officials called it an "isolated case." But though only a single cow was diagnosed, the discovery signaled the first real threat of the ravaging disease in North America. The announcement sent shock waves through the $80-billion-a-year beef industry, which fears the cattle cataclysm that shook the United Kingdom in the 1990s.

The occurrence of the scourge in North America is potentially "devastating," said John Lochte, executive director of R-CALF USA, a national advocacy group for cattle ranchers. "There's going to be a tsunami tomorrow."

No cases of mad cow disease have been found in the USA.

The United States is Canada's prime export market for beef. Canada sent 1.7 million heads of live cattle and 371,000 tons of beef product with a value of $2.5 billion to the USA last year.

Also last year, the United States produced 27.1 billion pounds of beef, the largest single sector of U.S. agriculture.

Mad cow disease infects humans as a variant of Creutzfeldt-Jakob Disease, which causes paralysis and death. In cattle, it is thought to spread through feed made with protein and bone meal from rendered cattle.

When doctors in Britain realized in 1996 that humans could get the brain-wasting disease from eating infected cattle, sales of beef in the U.K. plunged 40%, exports ceased and beef prices fell more than 25%. The economic loss to the country in the following year is estimated to be $1.2 billion to $1.6 billion.

The infected cow came from a farm in Alberta. It was slaughtered Jan. 31 and the carcass rendered into meat and bone meal, Canadian Agriculture Minister Lyle Vanclief said. Routine testing Friday turned up a possible case of mad cow, formally called bovine spongiform encephalopathy (BSE). The diagnosis was confirmed Tuesday morning.

No meat from the animal entered the human food chain, Vanclief said. Mad cow disease can be spread only by eating infected meat, especially infected brains and spinal cords.

Shares in fast food chains and meat producers and processors plummeted at the news, and cattle prices also fell.

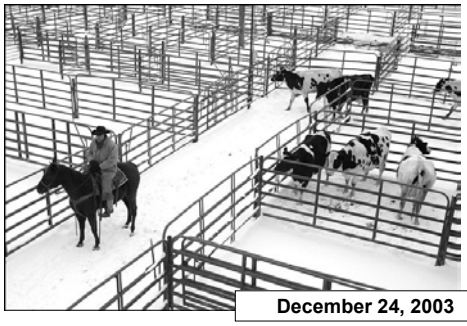
Agriculture Secretary Ann Veneman sought to reassure Americans that the "risk to human health and the possibility of transmission to animals in the United States is very low." I intend to eat a steak tonight," Veneman told CNN.

Contributing: Anita Manning, Patrick O'Driscoll, wire reports

U.S. beef industry gets a 'clarion call'
▸ Safety in focus, 3A
▸ Stocks fall, 18

May 23, 2003

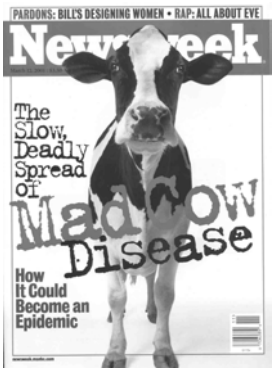
Empty cattle market in Washington



December 24, 2003

Chronology of BSE – US and Canada

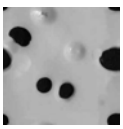
- *5/23/2003* - BSE diagnosed in cow in Alberta, US closed border
- *12/24/2003* - BSE diagnosed in cow in Washington State, Canadian origin
- *8/2004* -US opened borders to some Canadian beef, but no live cattle
- *12/29/2004* - US announced borders would reopen to live cattle in February of 2005
- *12/30/2004* – second BSE cow found in Alberta
- *1/11/2005* – third BSE cow found in Alberta

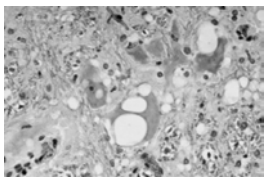


News

Harvard study finds BSE poses little threat to U.S. consumers, agriculture

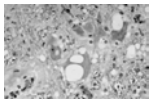
Still a need for added vigilance, according to Agriculture and Health and Human Services departments





BSE diagnosed in French goat, November 2004

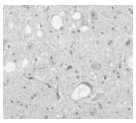
Feline spongiform encephalopathy



- >90 cases, primarily in UK

Chronic wasting disease

- A TSE
- First reported in **captive animals**, 1980
- Initially noted in MULE DEER

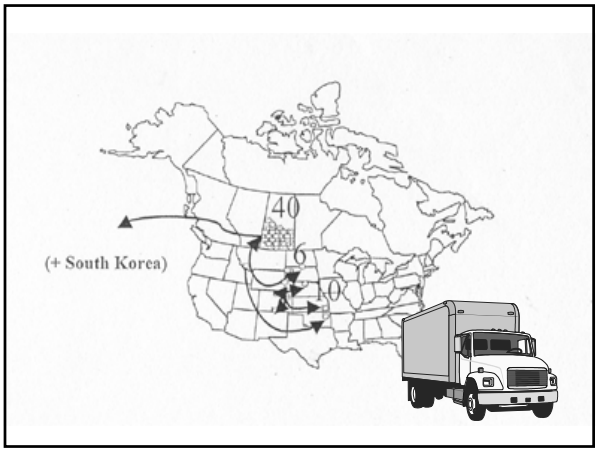


Chronic wasting disease

- Mule deer, white tailed deer, black-tailed deer, Rocky Mountain elk
- Poor body condition, excessive drinking and urination, repetitive actions, tremors, ataxia
- Many die of aspiration pneumonia

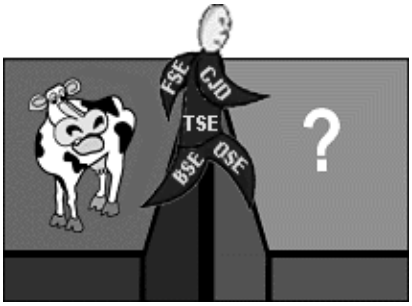
Chronic wasting disease

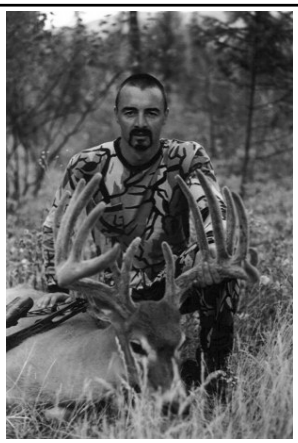
- Incubation period – not sure? 15 months?
- Experimental transmission by feeding – prions evident in tonsil and Peyer's patches



Chronic wasting disease
Transmission and Host Range

- Lateral transmission among cervids
- Intracerebral transmission to mice, ferrets, mink, goats, squirrel monkeys, calves
- No contact transmission from deer to cattle (yet)





Other 2005 Teleclasses

For more information, refer to www.webbertraining.com/schedule.cfm

- *February 24* - **Sneezes, Coughs and Drips: Respiratory and GI Outbreaks in Long Term Care** with Dr. Chesley Richards
- *March 10* - **Biocide Use in a Healthcare Environment** with Dr. Jean-Yves Mailard
- *March 17* - **WHO's Global Patient Safety Challenge 2005/2006 Preventing Healthcare Associated Infection; A Worldwide Strategy** with Dr. Didier Pittet
- *March 24* – **Infection Control in Pre-Hospital Care** with Margaret McKenzie
- *March 31* – **Voices of CHICA**
- *April 7* – **Root Cause Analysis for the Infection Control Professional** with Dr. Denise Murphy

Questions? Contact Paul Webber paul@webbertraining.com