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

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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
all muscle becomes meat

remainder is “rendered”

human consumption

meat and bone meal (animal feed)

HEAT

tallow

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BSE – A Zoonosis?

- March 20, 1996
- Cluster of unusual CJD cases

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

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<td>PrP plaques</td>
<td>frequent</td>
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153 cases, as of January 2005
16,700 appendectomy and tonsillectomy samples from 20-29 year olds
3 were positive, therefore 237 cases per million

Failure to recognize, failure to act…..

Will BSE continue to spread?
There is no live animal test.

Rapid Tests

- Western blot
- ELISA

Histopathology

Immunohistochemistry
Chronology of BSE – US and Canada

- 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

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

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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

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