Slide 1

Audio and Text:

Bovine Spongiform Encephalopathy
Basics of the Disease

Image:

None

Slide 2

Audio:
BSE is a progressive and fatal neurologic disease of cattle caused by an unconventional transmissible agent, the prion. BSE belongs to the family of diseases known as transmissible spongiform encephalopathies or "TSEs". In addition to BSE, TSEs include scrapie, which affects sheep and goats; transmissible mink encephalopathy; chronic wasting disease of deer and elk; and in humans, kuru, both classic and variant Creutzfeldt–Jakob disease or CJD, Gerstmann–Straussle–Scheinker syndrome, and fatal familial insomnia.

Text:
The Transmissible Spongiform
Encephalopathies (TSE's)

Image:
Two newspaper clippings.
First clipping:
Prion diseases in Animals. Scrapie in sheep has been known for centuries, but it doesn't seem to infect humans. Variants in other species have emerged more recently.
Scrapie, 1730s, sheep (photo of sheep)
Transmissible mink encephalopathy, 1965, minks (photo of mink)
Chronic wasting disease, 1980, elk and deer, (photo of elk and deer)
Bovine spongiform encephalopathy, 1985, cattle (photo of cow), 1990, cats (photo of cat), 1992, zoo monkey (photo of monkey).
Second clipping:
Prion Diseases in Humans
Creutzfeldt-Jakob: Begins subtly with depression and memory problems, but in 4 to 6 months progresses to dementia, uncontrollable jerking of muscles and finally death.
Gerstman-Straussler-Scheinker syndrome; Damage to motor portions of the brain makes walking, speaking and even swallowing difficult. Dementia comes much later. Death within 2-6 years.

Fatal insomnia: Progressive insomnia leads to panic attacks, phobias, hallucinations and finally dementia. Patients die within 18 months.
Kuru and variant Creutzfeldt-Jakob: Anxiety yields to trembling, loss of coordination and death after 16 months.

Slide 3

Audio:

The agents that cause BSE and other TSEs have yet to be fully characterized. The theory most widely accepted in the scientific community is that the agent is an abnormal prion protein. The BSE agent is extremely resistant to heat and to normal sterilization processes. It also does not evoke any detectable immune response or inflammatory reaction in host animals.

Text:

TSE's: Etiology

The leading theory is that TSEs are caused by a prion (proteinaceous infectious agent) capable of transforming normal body proteins to an abnormal form

Image:

Molecular illustration of a prion with a normal α-helix and the transformed abnormal B-sheet.

Slide 4

Audio:

Prions have many unique characteristics.

Text:

Prion

Host-coded normal cellular protein

Becomes partially protease resistant through a post-translational conformation change (alpha helix to beta sheet)

Smaller than smallest known virus

Elicits no detectable immune or inflammatory response in the host

Resistant to most disinfectants and treatments which normally destroy nucleic acids

Image:

None

Slide 5

Audio:

Cattle affected by BSE experience progressive degeneration of the nervous system.

Text:

Characteristics of TSE's

Brain degeneration with vacuolation

Long (2.5 years and >) incubation period

Infectivity very hard to inactivate

Progressive debilitating neurological

illness

Pathological changes

confined to CNS

Invariably fatal

Image:

Histologic slide of bovine brain tissue showing a spongiform change.

Slide 6

Audio:

Transmissible Spongiform Encephalopathies are so named because of the spongy appearance of the brain tissue in

infected animals when examined under a microscope.

Text:

None

Image:

Histologic slide of bovine brain tissue showing a spongiform change.

Questions:

1. Which of the following are characteristics of prions?

1) Their amino acid sequence is changed to make them protease resistant.

2) They are the same size as many viruses.

3) Post infection a mild inflammatory response may be detected in the host.

4) They are host-coded normal cellular proteins. (Correct)

5) Only disinfectants which destroy nucleic are effective against them.

2. TSE's generally have long incubation periods (2.5 years or greater).

1) True (Correct)

2) False

3. Pathological changes associated with the TSE's may be associated with multiple organ systems.

1) True

2) False (Correct)

Slide 7

Audio:

Affected animals may display nervousness or aggression, abnormal posture, difficulty in coordination and rising, decreased milk production, or loss of body weight despite continued appetite. All infected cattle die. There is neither any treatment nor a vaccine to prevent the disease.

Text:

BSE: Clinical signs

Changes in behavior

Hyperaesthesia

Weight loss with appetite retention

Drop in milk production

Ataxia

Difficulty in rising

Down or dead

Not all animals show all signs

Image:

Dairy cow with weight loss and wide based stance.

Slide 8

1. Which of the following is NOT a clinical sign of BSE?

1) Ataxia

2) Behavior changes

3) Hyperaesthesia

4) Increased appetite and weight gain (Correct)

5) Down or dead

Slide 9

Audio:

The incubation period from infection until first signs of disease, averages 3 to 6 years, although the period can be longer or shorter. Following the onset of clinical signs, the animal's condition deteriorates until it either dies or is destroyed. The process of deterioration usually takes from 2 weeks to 6 months.

BSE is not a contagious disease and therefore is not spread through casual contact between cattle. The primary source of BSE infection in cattle is commercial feed contaminated with the infectious agent. Scientific evidence shows that feed contamination results from incorporating ingredients

(for example, meat-and-bone meal) that contain protein derived from rendered infected cattle. Standard rendering processes do not completely inactivate or kill the BSE agent. Therefore, rendered protein such as meat-and-bone meal derived from infected animals may contain the infectious

agent. Regulations prohibiting the inclusion of mammalian or ruminant protein in ruminant feed, including cattle feed, are used to prevent BSE transmission.

Text:

BSE: Transmission

Contaminated feed

Clinical signs in 3-6 years

Image:

Beef cattle eating from a feed bunk.

Slide 10

Audio:

Consumption of feed contaminated with the BSE agent is the only documented route of field transmission of BSE.

Text:

BSE: Transmission

BSE spreads by:

Movement of incubating animals if incorporated into feed

Movement of BSE contaminated feed

Image:

Beef cattle eating from a feed bunk.

Slide 11

1. How is BSE transmitted?

Answer: Contaminated feed

Slide 12

Audio:

Research conducted to date indicates that only certain tissues in animals affected with BSE are actually infective. In cattle naturally infected with BSE, infectivity has been found only in brain tissue, the spinal cord, and the retina of the eye.

Text:

Distribution of infectivity:
natural cases

Image:

Illustration of a cow with the areas of the retina, brain, and spinal cord labeled.

Slide 13

Audio:

In experimentally infected cattle, the distal ileum of the small intestine, dorsal root ganglion, and trigeminal ganglion also were found to be infective. Some reports also include tonsils and bone marrow. The results on bone marrow, however have been inconclusive.

PI stand for post inoculation.

Text:

Distribution of infectivity:
experimental

Image:

Illustration of a cow with the areas of the brain (32 months PI), retina, trigeminal ganglia, spinal cord, dorsal root ganglia, distal ileum (6 months PI), and bone marrow labeled.

Slide 14

Audio:

Currently, there is no test to detect the disease in live cattle; veterinary pathologists confirm BSE by postmortem microscopic examination and/or the detection of abnormal prions in brain tissue.

Text:

Distribution of infectivity
High risk tissues for humans or animals – brain, spinal cord
Limits diagnostics – why there is no live animal test

Image:

Rancher touching the head of a beef cow.

Slide 15

Audio:

IHC stands for immunohistochemistry

WB stands for Western Blot

ELISA stands for Enzyme-linked immunosorbent assay

Text:

BSE: Diagnostics

No live animal test

No preclinical test

Need brain tissue

Histopathology

IHC, WB, ELISA

“Negatives” do not guarantee absence of infectivity

Image:

None

Questions:

1. Which of the following is NOT a location of BSE distribution?

1) Brain

2) Spinal cord

3) Cardiac muscle (Correct)

4) Retina

Questions:

1. As far as diagnostics, what is the major ramification of BSE being confined to the CNS?

Answer: No live animal test

Slide 16

Audio:

BSE was first diagnosed in 1986 in the United Kingdom. Since that time, it has been found in many European countries, and also in countries outside of Europe, including Japan, Canada, and the United States. To date, however, more than 95 percent of the total cases worldwide

have occurred in the United Kingdom.

Text:

Confirmed cases of BSE in Great Britain by year of detection (1987 – 2006)

Image:

Bar graph of the confirmed BSE cases in Great Britain by year of detection. The number of cases rise dramatically from almost none in 1987 to over 35,000 in 1992. The number of cases then drop dramatically to under 5000 by 1997. After 1997 the number of cases taper off to nearly 0 cases by 2006.

Slide 17

Audio:

This graph illustrates how the number of BSE cases decreased after the initial feed ban in 1988 and stopped after the complete feed ban in 1996.

Text:

UK: confirmed cases by year of birth

Image:

Bar graph of the confirmed BSE cases in Great Britain by year of birth. The number of cases rises quickly from almost no animals born before 1980 to over 35,000 cases of animals born in 1987. After the initial feed ban in 1988, the number of cases drops quickly to just over 5000 animals born in 1990. After 1990 the numbers taper off more gradually. In 1996 the complete feed ban was put into place and the case numbers born after that point dropped to 0.

Slide 18

Audio:

Over time many more countries detected BSE.

Text:

BSE Occurrence worldwide

Image:

Map of Europe and a timeline showing when the first case of BSE was detected in each country.

Ireland – Prior to 1986

Portugal, Switzerland – 1990

France – 1991

Netherlands, Belgium, Luxembourg – 1997

Denmark, Germany, Spain – 2000

Italy, Czech, Austria, Finland, Greece, Slovakia, Slovenia, Japan – 2001

Poland, Israel - 2002

Canada, USA - 2003

Slide 19

Audio:

Three cases of BSE have been detected in the United States.

The first BSE case was found in Washington State during December of 2003. It was found in a cow imported from Alberta, Canada and born in 1997.

The second BSE case was detected in a 12 year old Texas beef cow during June of 2005.

The third BSE case was detected in a 10 year old Alabama beef cow during March 2006.

Both indigenous US cases were born before the FDA feed ban of 1997.

Text:

BSE in the United States

Image:

Map of the United States with Washington State designated as December 2003, Texas State designated as June 2005, and Alabama State designated as March 2006.

Slide 20

Audio:

In 1996, scientists in the United Kingdom announced the finding of 10 cases of a new variant of Creutzfeld–Jakob disease or vCJD, a chronic and fatal neurodegenerative disease of humans. These new cases presented a different clinical and pathological picture from routinely diagnosed cases of classic or sporadic CJD. Experimental and epidemiologic studies have linked the occurrence of vCJD to exposure to the BSE agent, most likely through the consumption of food containing ingredients derived from infected cattle.

Text:

Why do we care about BSE?

Public Health Concern

Variant Creutzfelt-Jakobs Disease (vCJD)

Trade

What is the basis of the trade restrictions?

Image:

None

Slide 21

Audio:

There are several important differences between vCJD and CJD.

Text:

vCJD and CJD

vCJD is the human form of bovine spongiform encephalopathy

It is caused by eating beef infected with BSE

It occurs in young people

CJD is a similar neurologic disease

It is not caused by eating beef infected with BSE

It is seen in older people

Image:

None

Slide 22

Audio:

Three cases of vCJD have been reported from the United States. By convention, variant CJD cases are ascribed to the country of initial symptom onset, regardless of where the exposure occurred. There is strong evidence that suggests that two of the three cases were exposed to the BSE agent in the United Kingdom and that the third was exposed while living in Saudi Arabia.

Text:

Variant CJD case through February 2007

– total of 198 cases worldwide

Image:

Chart showing the vCJD cases by country through February 2007.

UK – 162 total primary cases with 6 alive, 3 secondary cases from blood transfusion with 1 alive, and 165 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

France – 21 total primary cases with 1 alive, 0 secondary cases from blood transfusion, and 1 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

Republic of Ireland – 4 total primary cases with 1 alive, 0 secondary cases from blood transfusion, and 2 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

Italy – 1 total primary cases with 0 alive, 0 secondary cases from blood transfusion, and 0 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

USA – 3 total primary cases with 0 alive, 0 secondary cases from blood transfusion, and 2 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

Canada – 1 total primary cases with 0 alive, 0 secondary cases from blood transfusion, and 1 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

Saudi Arabia – 1 total primary cases with 1 alive, 0 secondary cases from blood transfusion, and 0 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

Japan – 1 total primary cases with 0 alive, 0 secondary cases from blood transfusion, and 0 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

Netherlands – 2 total primary cases with 0 alive, 0 secondary cases from blood transfusion, and 0 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

Portugal – 1 total primary cases with 1 alive, 0 secondary cases from blood transfusion, and 0 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

Spain – 1 total primary cases with 0 alive, 0 secondary cases from blood transfusion, and 0 of these had a cumulative residence in the UK greater than 6 months during 1980-1996.

Slide 23

Audio:

Trade is a significant portion of the beef industry.

Text:

Trade

Approximately 9% of U.S. beef production is exported

Approximately 90% of the exports go to 4 countries (Japan, Korea, Mexico and Canada)

Image:

None

Slide 24

Audio:

Beef exports dropped significantly after BSE was detected in late 2003.

Text:

None

Image:

Bar graph showing US beef exports by country and carcass weight over time. It shows the carcass weight in million pounds increasing from 1000 to 2500 between 1990 and 2003. Of this the majority of exports were to Japan and South Korea. After 2003, the carcass weight in million pounds dropped to about 500 with no beef going to South Korea and Japan.

Slide 25

Audio:

The market losses associated with detecting BSE have been significant.

Text:

Losses

U.S. beef exports dropped 80% in 2004

Estimated loss approximately $3 billion (according to the National Cattleman's Beef Association)

Effects on consumer confidence

Image:

None

Questions:

1. BSE was first detected in the US in 2003. By what percentage did beef exports drop in 2004?

1) 50%

2) 60%

3) 70%

4) 80% (Correct)

5) 90%

Slide 26

Audio:

This concludes the course.

Text:

Thank-you!

www.aphis.usda.gov

Image:

Dairy cows.