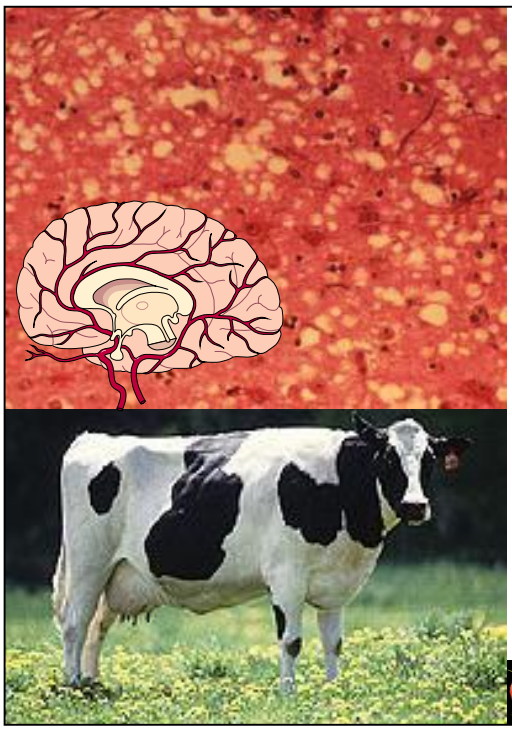


Hot Topics in Biology

Mad Cow Disease

By Sonia Clayton, PhD,
James Denk, MA, and
Nancy Moreno, PhD

BioEd Online

Image References:

Brain Tissue: Animal and Health Inspection Service. APHIS photos by Dr. Al Jenny

http://www.aphis.usda.gov/lpa/issues/bse/bse_photogallery.html Cow: US Food and Drug Administration.
http://www.fda.gov/oc/opacom/hottopics/bse.html (Accessed 1/14/04)

Cow:United States Department of Agriculture. Keith Weller

http://www.ars.usda.gov/is/graphics/photos/k5176-3.htm (accessed 1/14/04)

Brain:

Mad Cow Disease: An Introduction

- First identified by a veterinarian in England in 1986.
- Scientifically known as Bovine Spongiform Encephalopathy, or BSE.
- Disease that destroys portions of the nervous system in adult cattle.
- Name comes from the strange behavior and symptoms seen in cattle that have the disease.
- Disease may have passed to cattle through feed containing protein or bone meal from sheep infected with a similar disease.

Mad Cow Disease can be spread by animal feeds that contain protein supplements made from infected animals.

BioEd Online

Mad cow disease (known scientifically as Bovine Spongiform Encephalopathy, or BSE) is a disease that attacks the nervous system of adult cattle. The name "mad cow" comes from the strange behavior and symptoms seen in cattle that have the disease. These behaviors and symptoms include: aggression, lack of coordination with inability to stand or walk, and abnormal posture. BSE was first identified by a veterinarian in England in 1986. BSE is a fatal disease that progresses very slowly.

The name, Bovine Spongiform Encephalopathy, describes characteristics of the disease in scientific language. "Bovine" indicates that it is a disease of cattle. "Spongiform" refers to the spongy appearance of the brains of infected cattle. "Encephalopathy" is a term that refers to a disease of the brain, especially one involving alterations of brain structure.

Scientists believe the disease may have passed to cattle through feeds containing animal proteins from sheep with a similar disease, known as scrapie. The disease may spread to other cattle through feeds containing meat and bone meal from infected cows. In the United States, it has been illegal to feed most animal proteins to cattle since 1997.

References:

Brown P, Will RG, Bradley R, Asher DM, Detwiler L. 2001. Bovine spongiform encephalopathy and variant Creutzfeldt-Jakob disease: background, evolution, and current concerns. *Emerg Infect Dis* 7:6-16.

US Food and Drug Administration. http://www.fda.gov/oc/opacom/hottopics/bse.html (Accessed 1/14/04)

US Centers for Disease Control and Prevention. 2004. Bovine Spongiform Encephalopathy in a Dairy Cow -- Washington State. MMWR Weekly 52(53):1280-1285.

http://www.cdc.gov/mmwr/preview/mmwrhtml/mm5253a2.htm (Accessed 1/15/04)

Image Reference:

United States Department of Agriculture

http://www.ars.usda.gov/is/graphics/photos/animalsimages.new.htm (accessed 1/14/04)

Mad Cow Disease: Human Infection

- Scientists have linked Mad Cow Disease to a similar disease in humans.
- Humans contract the disease by consuming parts of the nervous system or meat contaminated by nervous system tissue from infected cattle.
- The human form of Mad Cow Disease is called variant Creutzfeldt-Jakob Disease (vCJD).

Cattle affected by Mad Cow Disease experience nervousness, aggression, or other changes in temperament; abnormal posture; lack of coordination; difficulty in rising; decreased milk production; and/or loss of weight despite continued appetite, followed by death.

APHIS photos by Dr. Art Davis

BioEd Online

The agent causing Mad Cow Disease has been linked through epidemiology studies (research that follows the causes and distribution of the disease among different populations) and laboratory research to a similar disease in humans that also damages that central nervous system. Scientists hypothesize that humans contract a form of Mad Cow Disease by consuming parts of the nervous system from infected cattle. The human form is called variant Creutzfeldt-Jakob Disease (vCJD), which is closely related to Creutzfeldt-Jakob Disease (CJD).

Refrences:

Brown P, Will RG, Bradley R, Asher DM, Detwiler L. 2001. Bovine spongiform encephalopathy and variant Creutzfeldt-Jakob disease: background, evolution, and current concerns. *Emerg Infect Dis* 7:6-16.

Centers for Disease Control and Prevention. http://www.cdc.gov/ncidod/diseases/cjd/cjd.htm. **(Accessed 1/14/04)**

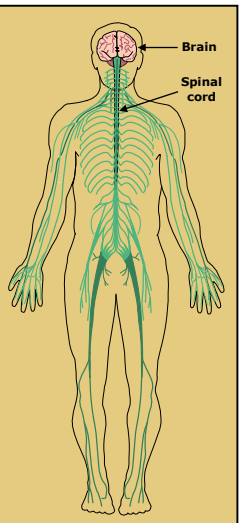
Image Reference**:**

Animal and Health Inspection Service

http://www.aphis.usda.gov/lpa/issues/bse/bse_photogallery.html

Variant Creutzfeldt-Jakob Disease (vCJD)

- vCJD is a fatal disease of the nervous system.
- vCJD is a form of Creutzfeldt-Jakob Disease (CJD), another fatal illness that affects the nervous system of humans.
- vCJD patients typically are much younger than CJD patients. In the United States, vCJD has been observed to appear at a median age of 26 years, while CJD appears at 68 years.
- vCJD can be confirmed only through examination of the patient's brain tissue (through biopsy or autopsy).



Creutzfeldt-Jakob Disease attacks the central nervous system, which consists of the brain and spinal cord.

BioEd Online

Variant Creutzfeldt-Jakob Disease (vCJD), is a fatal disease of the nervous system in humans. In the United States,the average age of patients with vCJD is 26 years, as opposed to 68 years for those who contract Creutzfeldt-Jakob Disease (CJD). This age discrepancy led researchers to look carefully at the younger patients and to uncover the connection between vCJD and Mad Cow Disease. On the basis of observable symptoms, probable cases can be diagnosed. However, confirmation of vCJD cases requires examination of patients' brain tissue after death.

References:

Centers for Disease Control and Prevention. http://www.cdc.gov/ncidod/diseases/cjd/cjd.htm. **(Accessed 1/14/04)**

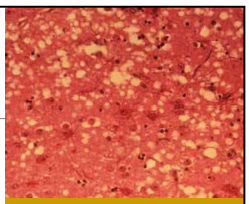
Department of Health and Human Services. Center for Disease Control and Prevention. Morbidity and Mortality Weekly Report 2002; 51(41): 927-929. http://www.cdc.gov/mmwr/preview/mmwrhtml/mm5141a3.htm (Accessed 1/16/04)

Hill AF, Zeidler M, Ironside J, Collinge J. 1997. Diagnosis of new variant

Creutzfeldt-Jakob disease by tonsil biopsy. *Lancet* 349:99-100.

Mad Cow Disease: Transmission

- Scientists believe that Mad Cow and related diseases are caused by abnormal proteins in the nervous system, called prions.
- Prions are different from bacteria, viruses or any other known disease-causing agent.
- Prions enter the body when people eat infected beef products.
- Once in the body, prions are believed to convert normal proteins in the nervous system into the abnormal prion shape.
- Cumulative damage by prions leads to Mad Cow and other similar diseases of the brain and nervous system.



The brains of affected cows have a sponge-like appearance when tissue sections are examined in the lab.

APHIS photos by Dr. Al Jenny

BioEd Online

Prion diseases attack nervous system tissue. Prions have been associated with Bovine Spongiform Encephalopathy (BSE), Creutzfeldt-Jakob Disease (CJD), and variant Creutzfeldt-Jakob Disease (vCJD). Neither viruses nor bacteria, prions are thought to be proteins with abnormal shapes. Prions enter the body when people eat beef products that contain infected nervous system tissue. Once in the body, prions are believed to convert normal proteins in the nervous system into the abnormal prion shape.

Prions duplicate by attaching themselves to, and changing the structure of, normal proteins. It is believed that when a person eats the brain, spinal cord or other nervous system tissue from an infected animal, prions are absorbed into the body, where they slowly begin to transform normal proteins, eventually leading to fatal damage to the nervous system.

References:

Nunziante M, Gileh S, Schatzl HM. 2003. Prion Diseases: from molecular biology to intervention strategies. *Chembiochem* 4(12): 1268-1284.

Mallucci G, *et al*. 2003. Depleting Neuronal PrP in prion infection prevents

disease and reverses spongiosis. *Science* 302:871-874.

Cohe, FE, Pan KM, Huang Z, Baldwin M, Fletterick RJ, Prusiner SB. 1994. Structural clues to prion replication. *Science* 264:530-531.

Si K, Lindquist S, Kandel ER. 2003. A neuronal isoform of the aplysia CPEB has prion-like properties. *Cell* 115(7):879-891.

U.S. Food and Drug Administration, Center for Food Safety and Applied Nutrition, *Foodborne Pathogenic Microorganisms and Natural Toxins Handbook*. http://vm.cfsan.fda.gov/~mow/prion.html. **(Accessed 1/14/04)**

Image Reference:

Animal and Health Inspection Service

http://www.aphis.usda.gov/lpa/issues/bse/bse_photogallery.html Cow: US Food and Drug Administration. http://www.fda.gov/oc/opacom/hottopics/bse.html (Accessed 1/14/04)