Bovine Spongiform Encephalopathy Fact Sheet

1. **What is Bovine Spongiform Encephalopathy (BSE)?**

a. BSE, often called "Mad Cow Disease," is a fatal prion disease affecting the brain and spinal cord of cattle. The word "prion" is an abbreviation for "infectious protein."

b. Prions are abnormal variants of proteins that occur normally in cells, such as human brain cells. Amazingly, when prions enter the body, they are able to convert their normal counterparts into more of the abnormal forms.

c. The difference between the normal and abnormal proteins does not lie in their primary structure (the sequence of their amino acids), but rather in their 3-dimensional folding. The abnormal intracellular PrP proteins are folded in a way that allows them to resist normal protease degradation so that over time this leads to the build-up of aggregates of prions in the neurons in the brain.

d. Creutzfeldt-Jacob Disease (CJD) is a human prion disease. In both CJD and variant CJD (vCJD), prions accumulate in the brain cells causing progressive brain damage, gait and balance disturbances, difficulty swallowing, weight loss, behavioral changes and eventually death.

e. When viewed at autopsy, stained tissue slides from the brains of animals or humans with prion disease are filled with characteristic microscopic holes created by the excess accumulation of abnormal prion proteins in the cells of the brain.

2. **Where does BSE occur?** - The vast majority of cattle with BSE have been reported from Europe, especially the United Kingdom (U.K.). Human cases of vCJD resulted when people ate infected parts of cattle with BSE. Most of the cases of this human disease were reported from the U.K. and Europe (http://www.cdc.gov/ncidod/dvrd/vcjd/index.htm).

a. Through the end of 2012, more than 183,317 cases of BSE were confirmed in the United Kingdom in more than 36,194 cattle herds.

b. The BSE epidemic in the U.K. peaked in January, 1993, at almost 1,000 new cases in cattle per week. Through measures taken to protect the cattle feed supply, disease has declined since then. Only 69 cases of BSE in cattle have been diagnosed in the U.K. since 2007, with only two of these in 2012.

c. The disease outbreak most probably originally resulted from feeding cattle with meat-and-bone meal from sheep with a different type of prion disease called Scrapie. There is strong evidence that the BSE outbreak was then amplified by feeding infected cattle derived meat-and-bone meal to young calves as a feed supplement.

3. **Is BSE occurring in the United States?** - The United States Department of Agriculture (USDA) conducts special surveillance for BSE in cattle in the United States:

a. The first known case of BSE in the United States was identified in December, 2003, when the U.S. Department of Agriculture (USDA) announced a presumptive diagnosis of BSE in an adult Holstein cow from Washington State. Investigation revealed that the cow was imported into the United States from Canada in August, 2001.

b. Three more cattle have been diagnosed with atypical BSE (see below) in the United States.

(1) In 2005, a second case of BSE was confirmed in a Texas cow, bred and raised in the United States.

(2) The third case was in 2006 in an animal in Alabama whose origin could not be established.

(3) In May 2012, the fourth case of BSE was confirmed in a California cow, bred and raised in the United States.

4. **Is BSE a food borne hazard in the United States?**

a. BSE has been transmitted to humans primarily in the United Kingdom, causing vCJD. In the U.K., where over 1 million cattle have been infected with BSE and many were consumed by humans, a substantial species barrier (a natural resistance to spreading prion infection between different species of animals) appears to protect humans from widespread illness. Prior to 2013,, a total of 225 cases of vCJD had been reported worldwide; of these, 176 had occurred in the U.K.

b. The Classic (C-type) prion phenotype is the cause of the BSE and vCJD outbreaks in the U.K. Classic BSE usually strikes cattle between 4 and 8 years of age.

c. Atypical BSE is characterized by a higher (H-type BSE) or lower (L-type BSE) molecular mass Prion protein than the form that occurs in classical BSE. Unlike classical BSE, atypical BSE has been detected mainly in older cattle which suggest they are a sporadic form of BSE.

d. All four U.S. BSE cases were caused by the atypical prion. Only 60 cases of atypical BSE have been identified worldwide. No humans are known to have contacted vCJD from cattle infected with atypical BSE.

5. **Is there evidence directly linking vCJD to BSE exposure**?

a. There is strong epidemiologic and laboratory evidence for a causal association between vCJD and BSE. The absence of confirmed cases of vCJD in geographic areas free of BSE supports a causal association. In addition, exposure of the population to potentially BSE-contaminated food (1984-1986) and onset of initial vCJD cases (1994-1996) is consistent with the incubation periods for the typical form of CJD.

b. Recent declines in the occurrence of vCJD human disease in the U.K. coincide with declines in the incidence of BSE in cattle.

6. **Are increased surveillance efforts in place to determine whether vCJD occurs in the United States?**

a. Yes. In addition to ongoing review of national CJD mortality data, the Centers for Disease Control and Prevention (CDC) is conducting active CJD surveillance at its Emerging Infections Program Monitoring Sites.

b. In 1997, CDC, in collaboration with the American Association of Neuropathologists, established the National Prion Disease Pathology Surveillance Center at Case Western Reserve University, Cleveland, Ohio. This pathology center provides free, state-of-the-art diagnostic services to United States physicians. It also helps to monitor the possible occurrence of emerging forms of prion diseases, such as vCJD, in the United States. See its website at: http://www.cjdsurveillance.com.

7. **Has there ever been a human vCJD case in the United States?**

a. In 2002, CDC reported a case of vCJD in a Florida resident who was born in and grew up in the United Kingdom during the BSE epidemic.

b. Another human case of vCJD was reported in 2005 in a man originally from the U.K. who lived in Texas for 4 years.

c. A third patient with vCJD was reported in 2006. The patient was born and raised in Saudi Arabia and had only permanently moved to the US a year earlier. The patient was likely infected as a child while living in Saudi Arabia.

d. In all three cases, none are thought to have any health implications for the United States.

8. **Is BSE a food borne hazard for travelers to Europe?** - The current risk for infection with the BSE agent among travelers to Europe is extremely small, if it exists any longer at all.

9. **For more information:** http://www.cdc.gov/ncidod/dvrd/bse/index.htm

This fact sheet provides general information. Please contact your physician and/or veterinarian for specific clinical information related to you or your animal.