**Bovine Spongiform Encephalopathy**

**Classic Case:** Adult cattle, insidious onset of abnormal behavior, progressive over 1-6 months

**Presentation:**

- Abnormal behavior +/- CNS signs
  - Apprehensive, aggressive
  - Hyperesthesia
  - Ataxia, incoordination
  - Difficulty negotiating obstacles
  - Hypermetria, tremors
  - Low head carriage
- Weight loss
- Resistance to milking
- Reduced milk yield
- Bradycardia
- Reduced ruminal activity
- No Pruritis ★

★ ★ **BSE is a REPORTABLE ZOONOTIC disease ★ ★

**DDX:** Meningitis Rabies Listeriosis
Lead poisoning Ketonuria Brain tumor
Brain abscess Hypomagnesemia Polioencephalomalacia
Hepatic encephalopathy Organophosphate toxicity

**Test of choice:** must identify presence of abnormal prion protein

- No antemortem tests; freezing of post mortem tissues is OK
- Screening test for BSE - ELISA – on the **OBEX** area of brainstem
- Inconclusive results are sent to NVSL for confirmation
  - Approved labs - National Animal Health Laboratory Network (NAHLN)
  - USDA’s National Veterinary Services Laboratories (NVSL)
- Confirmatory tests
  - **Immunohistochemistry** of OBEX +/- electron microscopy
    - Also provides visual identification of tissue damage in brain
    - Vacuoles, protein accumulation in brain
  - Western blot - for autolyzed/degraded samples

**No serology for BSE or other TSE’s –**
No immune response, No antibodies produced

*OBEX – in brainstem where 4th ventricle narrows caudally; Image courtesy of CDC*
Rx of choice:

No treatment
Euthanasia advised as soon as Dx is confirmed

Prognosis:

Grave; Fatal disease

Prevention:

- **Do not feed animal tissues/products to cattle**
  - 1997 - Mammalian proteins banned from ruminant feed
  - ‘High risk’ tissues banned for all animal feed since 2009
- Best method to destroy prion – incinerate carcasses
- Use only disinfectants approved for prions
- Always consider BSE in ‘downer’ cows
- **Take GREAT care - handling or transporting tissues**

Pearls: **“MAD COW DISEASE”**

| First recognized in the United Kingdom 1986 | Canada – 1<sup>st</sup> case -1993, cow from UK |
| ~200,000 cases in bovine; 97% in the UK | US - 3 cases 2003 - 2006 |
| Became a REPORTABLE disease in 1988 | 4<sup>th</sup> case April 2012 |
| APHIS tests 40,000 cattle per year in ongoing surveillance program | |

BSE is a transmissible spongiform encephalopathy (TSE)

- TSEs are caused by prion proteins - **Prions (PrP)**
  - Altered form of a normal protein
  - Same primary a.a. sequence
  - Secondary structure – ‘misfolded’
  - Protease & disinfectant resistant
  - Prions pseudo-'reproduction' - convert normal proteins to PrP’s
- Spread by ingestion
- Infected cattle - no symptoms for years
- No genetic susceptibility
- PrP not ‘foreign’ – *no immune response*
- USDA tests 40,000 cattle each year in ongoing surveillance program
**Zoonotic:** BSE in people IS the variant form of Creutzfeldt-Jakob disease (vCJD)

**Images/Links worth a look:**

- **BSE Powerpoint** with notes, Center for Food Security and Public Health, Iowa State University
- **Normal protein structure review**, and changes seen with PrP diseases, from Arizona biology students
- **APHIS BSE website**, **Confirmatory Dx tests for BSE**, **BSE at CDC** (includes good historical info)
- **Normal vs Abnormal PrP's**

![Spongy appearance of brain in cow with BSE; (courtesy USDA)](image)


**My Notes:**