**Bovine Spongiform Encephalopathy (BSE)**

**Extended version**

**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
- Lead poisoning
- Brain abscess
- Hepatic encephalopathy
- Rabies
- Ketosis
- Listeriosis
- Brain tumor
- Hypomagnesemia
- Polioencephalomalacia
- 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

No immune response, so no antibodies produced

**OBEX** – in brainstem where 4th ventricle narrows caudally; Image courtesy of CDC
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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\textsuperscript{st} 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\textsuperscript{th} 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
- 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