



# Bovine Spongiform Encephalopathy

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 ★



Downer Holstein with BSE  
(courtesy USDA)

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

## DDX: Meningitis

Lead poisoning  
Brain abscess  
Hepatic encephalopathy

Rabies

Ketosis  
Hypomagnesemia  
Organophosphate toxicity

Listeriosis

Brain tumor  
Polioencephalomalacia

**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



OBEX – in brainstem where  
4<sup>th</sup> ventricle narrows caudally;  
Image courtesy of CDC

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

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## Rx of choice:

No treatment  
Euthanasia advised as soon as Dx is confirmed

## Prognosis:

Grave; Fatal disease



*Holstein with BSE (courtesy CDC)*

## 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  
~200,00 cases in bovine; 97% in the UK  
Became a REPORTABLE disease in 1988  
APHIS tests 40,000 cattle per year in ongoing surveillance program

Canada – 1<sup>st</sup> case -1993, cow from UK  
US - 3 cases 2003 - 2006  
4<sup>th</sup> case *April 2012*

**BSE is a transmissible spongiform encephalopathy (TSE)**

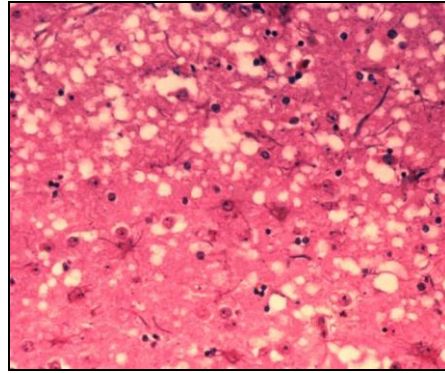
- TSEs are caused by prion proteins - **Prions (PrP)**
  - Altered form of a normal protein
  - Same primary a.a. sequence
  - Secondary structure – 'misfolded'
  - Protease & disinfectant resistant
  - Prions pseudo-'reproduction' - convert normal proteins to PrP's
- Spread by ingestion
- Infected cattle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



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**Zoonotic:** BSE in people IS the variant form of Creutzfeldt-Jakob disease (vCJD)



*Spongy appearance of brain  
in cow with BSE; (courtesy USDA)*

**Refs:** LA Neurology, Mayhew, 2<sup>nd</sup> ed. p. 259, and BSE from Merck Vet Manual, 10<sup>th</sup> ed., online.

**My Notes:**