

# Food Safety

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A review on how BSE (Bovine Spongiform Encephalopathy) can cause

CJD (Cruitzfeldt Jacob Disease)

Food intoxication

## How does this intoxication occur

- BSE infected beef
- Ingestion of the beef by human
- Contraction of CJD
- Intoxication and death

## What is BSE

A disease which attack Central Nervous System of cow which can be transmitted to human by consumption of the infected beef ■

Causing numerous hole in the brain when under microscope ■

Giving spongy appearance ■

## How did it happen first

First reported by Colin Whitaker , a veterinary surgeon in England in 1986 ■

Diagnosis was : ■  
hyperactivity  
aggressiveness towards the farmer  
ataxia ( incoordination)

A symptom resembling to deficiency of calcium in diet or organophosphate intoxication, which both were ruled out

## Spread of BSE

- The first case did not seem to be important
- But occurrence of the next and the next and so on until, by the end of 1986 nine herds had been affected
- The incidence of the disease increased dramatically until its peak (7267 herds affected) in 1992
- Following that event the worst scenario came up
- And that was intoxication of the beef consumers and eventually death

## Other kinds of BSE

- Indicative of a group of diseases called TSEs (Transmissible Spongiform Encephalopathy)
- Transmissible: can be transferred
- Spongiform: because of the spongy microscopical appearance of the brain
- Encephalopathy: disease of the brain
- Therefore **TSE**

## Other animal TSEs

- Scrapie: in sheep and goats  
The first TSE discovered reported in 1732, but until the early part of the 20<sup>th</sup> century was rarely considered
- Human TSE “CJD” :  
first reported in 1920 by Hans Cruitzfeldt  
The following year Alfred Jacob reported 4 cases
- The disease now bears both names in recognition of their contribution **CJD**

## What is CJD

- The clinical cause and features of CJD is have been well documented
- A rare disease
- Found world wide at a rate of approximately 1-2 per million per year
- Has the same symptom of BSE

## What causes BSE

Works of Professor Stanly Prusiner ■  
on scrapie revealed the nature of  
BSE

The agent is not a bacterium ■

Not a virus either ■

So what IS IT ?????????????? ■

## A damaged Prion

Professor Prusiner discovered a truly ■  
amazing causative agent  
Called it a proteinaceous infectious particle ■  
called Prion

It is a protein ■

It is not alive ■

But behaves like any other infectious ■  
agent

It replicate itself in an infected animal ■  
body

## Recognition of Prion as a causative agent

- The theory of infectious Prion was not simply accepted by other scientists
- However the Prion stood the test of time and incredible scientific scrutiny
- Now it is accepted as the causative agent of TSE
- Prusiner was awarded Noble Prize in 1997 for his discovery

## What is Prion

- Prions are medium sized protein (MW=33-35000 Dalt)
- Found in most animal cell especially in the brain
- Play a roll in communication (recognition between cells)
- Called cellular Prion protein "*PrP<sup>C</sup>*" (*Pr=Prion, P=Protein C=cellular*)

## Infectious *Prion*

- In contrast there are damaged form of proteins which resemble "*Pr<sup>PC</sup>*" very closely
- But different enough not to function properly
- So called Scrapie Prion "*Pr<sup>PSC</sup>*"
- It has got "s" because first was identified from scrapie sheep

## "*Pr<sup>PC</sup>*" versus "*Pr<sup>PSC</sup>*"

- They have both the same amino acid building blocks
- They have the same MW
- But important difference: the shape of protein make up is different, ie they different in quaternary structure,
- of in another word folding (conformation) "*Pr<sup>PSC</sup>*" and "*Pr<sup>PC</sup>*" is different
- "*Pr<sup>PC</sup>*" has a lot of alfa helix while "*Pr<sup>PSC</sup>*" has more beta pleated sheet

## Similarity of "*PrP<sup>SC</sup>*" and *CJD Prion*

- Human get CJD, is this the human form of scrapie????????? ■
- Studied showed that they not connected ■
- CJD caused by spontaneous change in gene (mutation) that codes for human "*PrP<sup>C</sup>*" resulting in synthesis of *CJD Prion* rather like "*PrP<sup>SC</sup>*" that has the same devastating effect ■

## So how does BSE cause CJD?

- Researches had shown that a new form of CJD, which they called a new variant CJD (*nvCJD*) was caused by eating BSE infected beef ■

## So the big question *what is nvCJD or vCJD*

- Disease caused by eating BSE infected beef
- First appeared in England in 1995/6
- The cases were very like CJD
- But CJD occurs in late life normally after 50s
- While vCJD occurs in their teen and 20s
- CJD period to death is 4 months
- vCJD period to death is 14 months

## How does the Prion get to the brain

- Absorption from the GI
- Travel to spinal cord
- Slow movement towards the brain (the process is by diffusion therefore slow process)
- Incubation period about 10 years  
(Remember occurrence of BSE in 1996, and appearance of vCJD in 1995/6)
- *Contact with native "Pr<sup>PC</sup>"*
- Conversion of native "Pr<sup>PC</sup>" to "Pr<sup>PSC</sup>"
- Then death is inevitable

## The origin of BSE

- Why was the first case in 1986???
- Prions are passed from animal to animal orally
- In cattle breeding MBM used as a component of cattle feed, a way of putting back valuable elements into farm animals' food chains
- In 1980s talow extraction was changed from solvent to heat
- It is thought that this process caused survival of BSE Prion and infect cattle via the infected MBM

## Another explanations

- It is thought that scrapie Prion has jumped the species barrier and infected cows through sheep MBM used to feed cattle
- A better and now widely accepted explanation: "*a Prion mutant cow*" made into MBM
- *This can happen anywhere which might explain the few BSE cases around the world*

## Conclusion

### how to prevent contracting this deadly disease?

- *PrP<sup>SC</sup>* heat stable
- *PrP<sup>SC</sup>* withstand normal metabolism of protein in animal body
- Therefore prevention of occurrence of the disease and
- Avoiding consumption of infected beef if occurred is the key factor
- Therefore, tight surveillance of cattle breeding and detection of any case if happen
- control over MBM produced from sheep and cows
- Banning of consumption of beef especially offals if any case detected
- Probable use of medicine to inactivate *PrP<sup>SC</sup>*

**Thank you**

**keep smiling**

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