

# Bovine Spongiform Encephalopathy

## How does it cross the species barrier ?



# Transmissible Spongiform Encephalopathies (TSEs)

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## *Human diseases*

### *Creutzfeldt-Jakob Disease*

- *sporadic*
- *iatrogenic*
- *familial*
- *«variant»*

### *Kuru*

### *Gerstmann-Sträussler-Scheinker*

### *Fatal Familial Insomnia (FFI)*

## *Animal diseases*

### *Scrapie of sheep and goat*

### *Bovine Spongiform Encephalopathy*

### *Chronic Wasting Disease of deer and elk*

### *Transmissible Mink Encephalopathy*

### *Feline Spongiform Encephalopathy*

# Characteristics of TSEs

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**Transmissible neurodegenerative diseases**

**Long asymptomatic incubation period**

**Fatal**

**Neuropathology : neuronal death, spongiosis, gliosis**

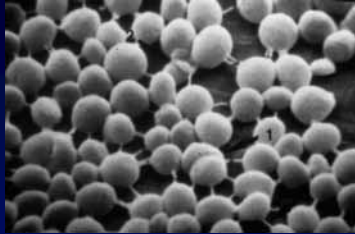
**Absence of systemic inflammation or specific immune response**

**Absence of prophylaxis or treatment**

**Etiology? Non Conventional Transmissible Agents (NCTA) or Prions**

**Infectious titres :  $10^{10}$  IU/g of brain**

Bacteria



Virus



Infectious  
agent ?

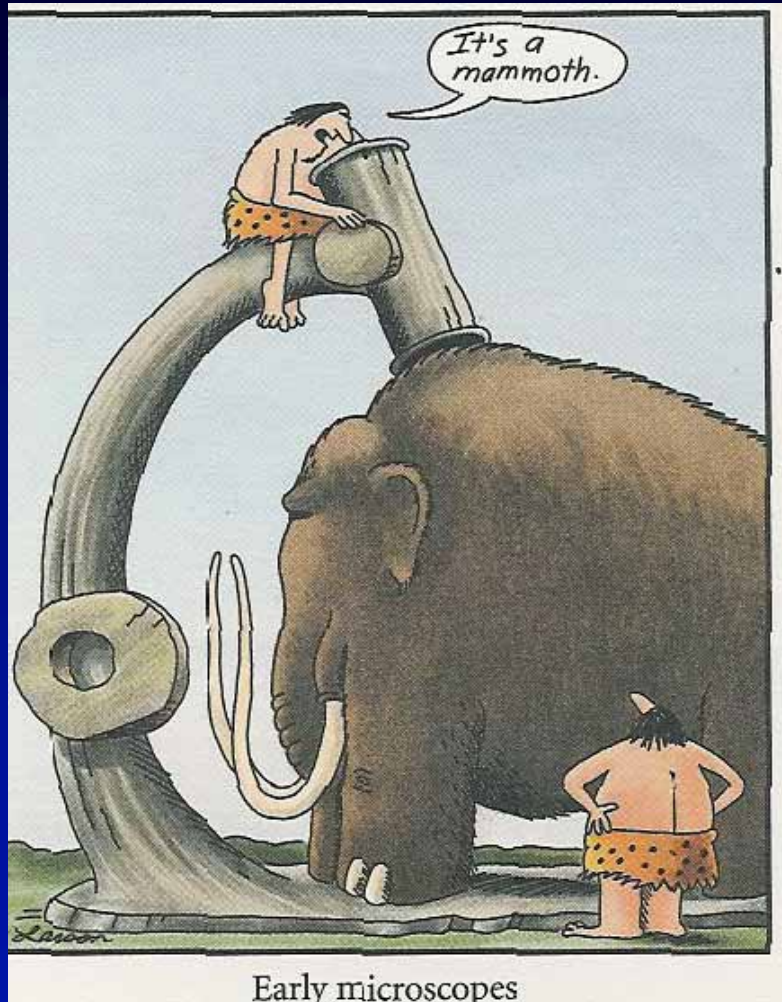
Protozoa



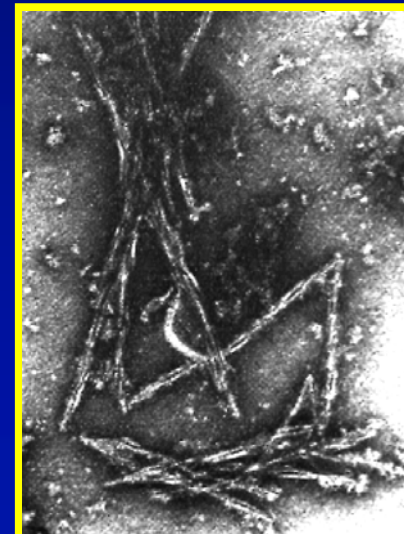
Fungi



# The search of the infectious particle



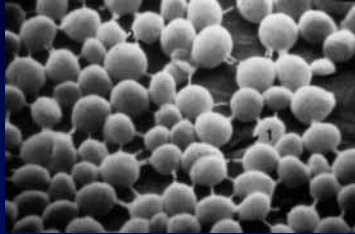
Amyloid deposit



proteinaceous infectious  
particle  
(Prion)

SAF (P. Merz, 1981)

Bacteria



Virus



Protein !!

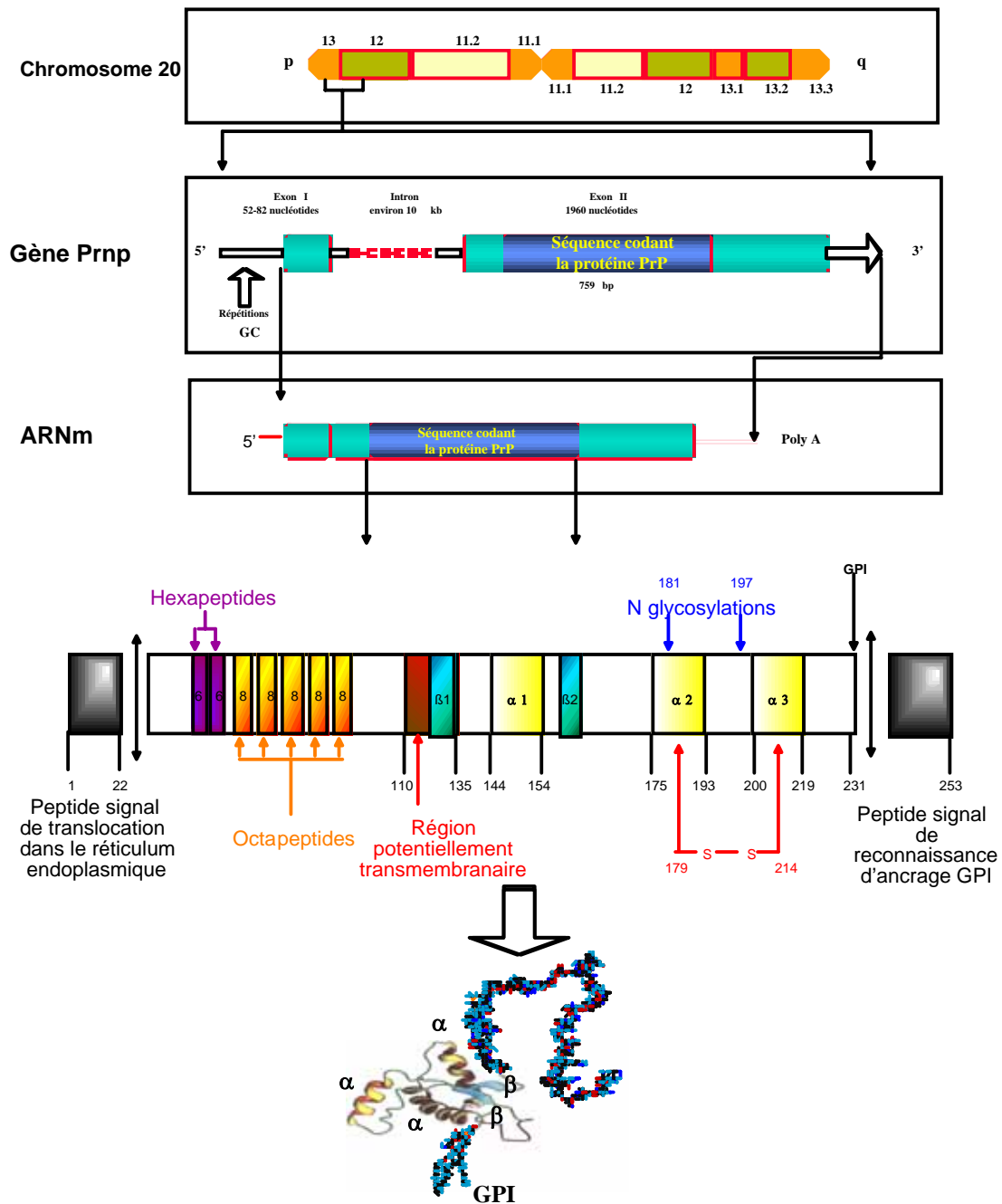
Protozoa



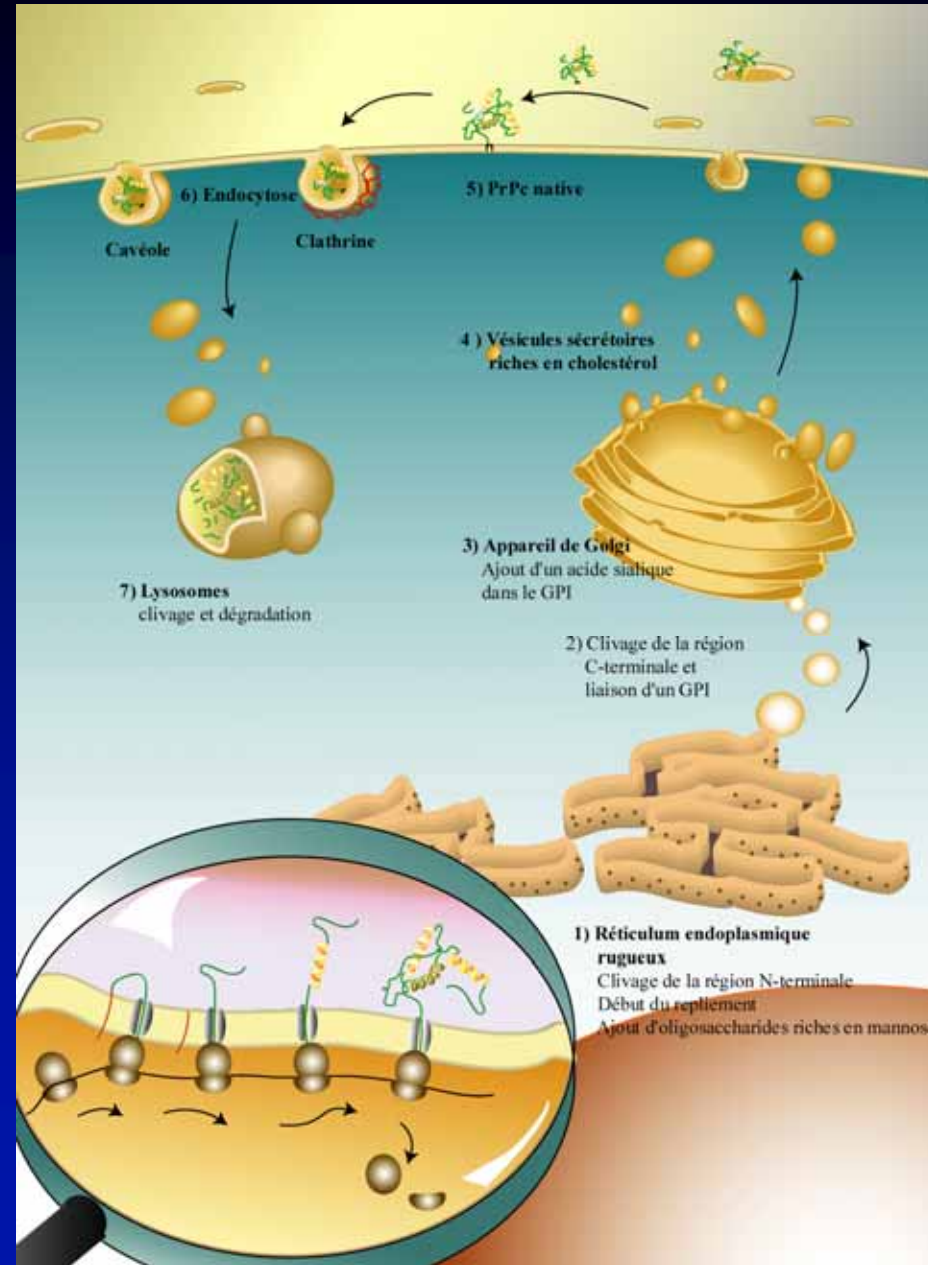
Fungi



*The prion protein is synthesized by the host*



# Cellular cycle of the prion protein



# How can a host protein be infectious ?

Two Forms



of the same protein ?



# How can a host protein be infectious ?

Two Forms



of the same protein ?

Normal brain



+

Protease

Sensitive

~~Cellular PrP~~

~~PrP<sup>C</sup>~~



# How can a host protein be infectious ?

Two Forms

of the same protein ?



Normal brain

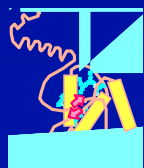
Scrapie infected brain

■  
+  
Protease

Sensitive

Partially resistant

~~Cellular PrP<sup>C</sup>~~  
~~PrP<sup>C</sup>~~



Scrapie PrP<sup>Sc</sup>

# Diagnosis of prion diseases

## Only specific biochemical marker : PrPres

PrP<sub>c</sub> : cellular prion protein (normal form)

PrP<sup>res</sup> : abnormal form of PrP, co-purifies with infectivity, resistant to proteases

