

Bovine Spongiform Encephalopathy
How does it affect the safety of beef ?

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BSE (牛海綿状脳症)
BSEと牛肉の安全性

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Transmissible Spongiform Encephalopathies (TSEs)

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

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伝達性海綿状脳症 (TSEs)

ヒトの疾病

CJD

- 孤発性
- 医原性
- 家族性
- 変異型

クールー病

ゲルストマン・シュトライスラー・シャインカー

致死性家族性不眠症 (FFI)

家畜の疾病

めん羊・山羊のスクレイピー

BSE (牛海綿状脳症)

シカやエルクの慢性消耗病 CWD

伝達性ミンク脳症 (TME)

猫海綿状脳症

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Characteristics of TSEs

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

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TSEsの特徴

伝達性神経変性疾病

長い無症状潜伏期間

致死性

神経病理学:ニューロン(神経細胞)死、海綿状、グリオシス

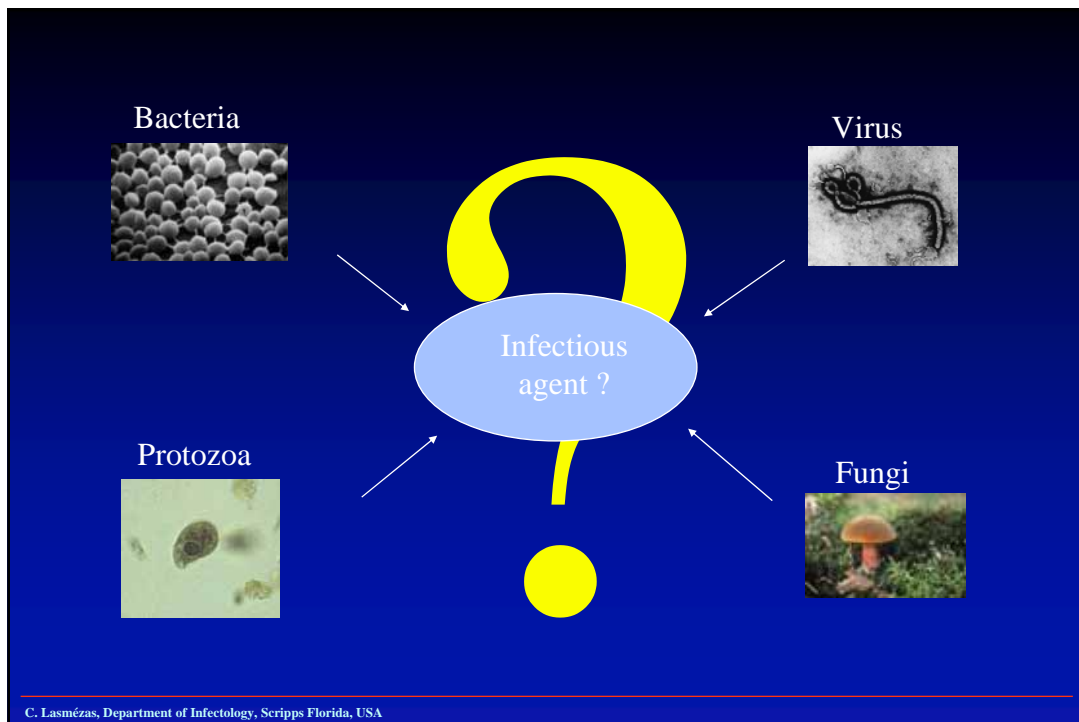
全身性炎症や特定の免疫反応がないこと

予防や治療法がないこと

病因学? 特殊伝達病因(NCTA)ないしはプリオン

感染価:脳 10^{10} IU/g

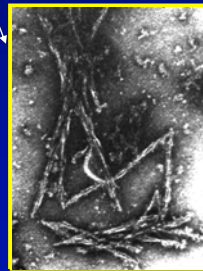
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The search of the infectious particle



Amyloid deposit

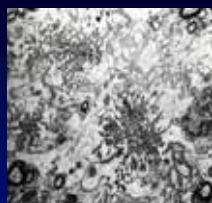
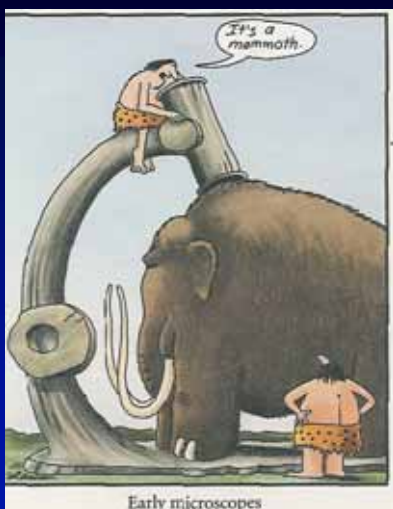


= proteinaceous
infectious
particle
(Prion)

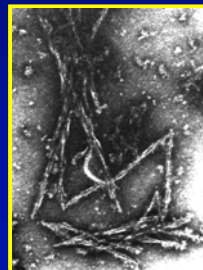
SAF (P. Merz, 1981)

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感染性粒子の探索



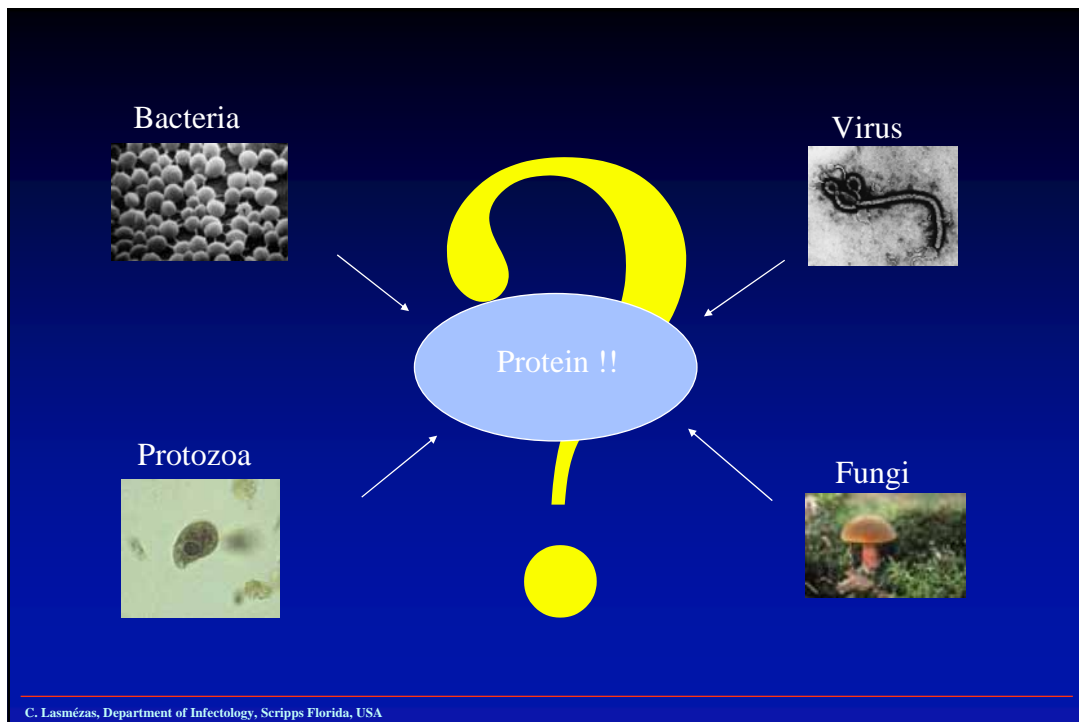
アミロイド沈着



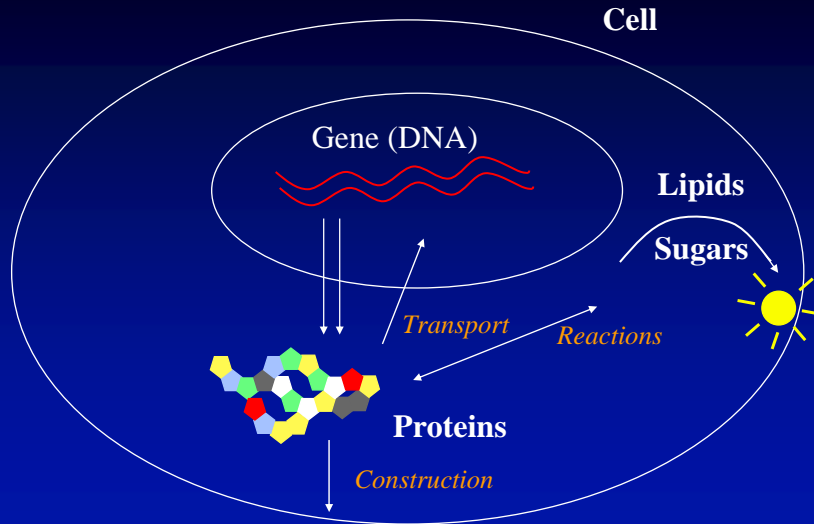
= たん白性感染性粒子
(プリオン)

SAF (P. Merz, 1981)

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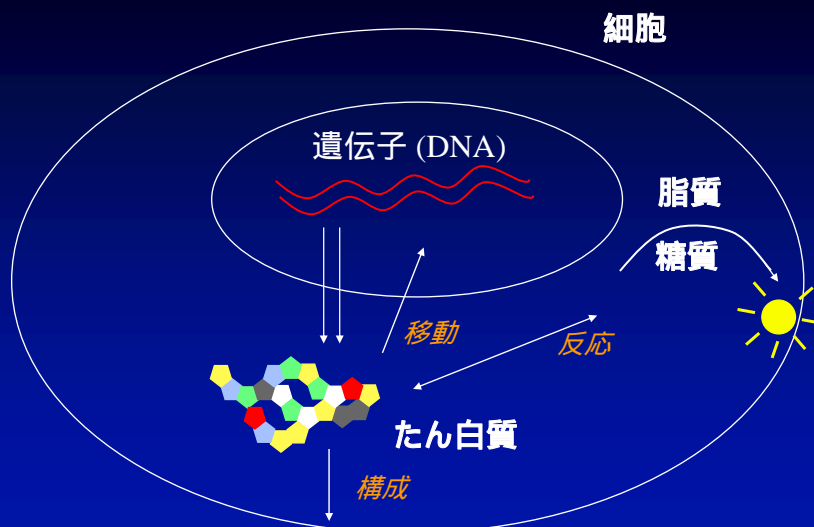


What is a protein ?



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たん白質って何？



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How can a host protein be infectious ?

Two Forms



of the same protein ?

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どのようにして宿主たん白質は感染可能となるのか？

同じたん白質の



二つの型？

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How can a host protein be infectious ?

Two Forms



of the same protein ?

Normal brain

+
Protease

Sensitive

~~Cellular PrP~~
~~PrP^C~~



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どのようにして宿主たん白質は感染可能となるのか？

同じたん白質の



二つの型？

正常な脳

+
プロテアーゼ

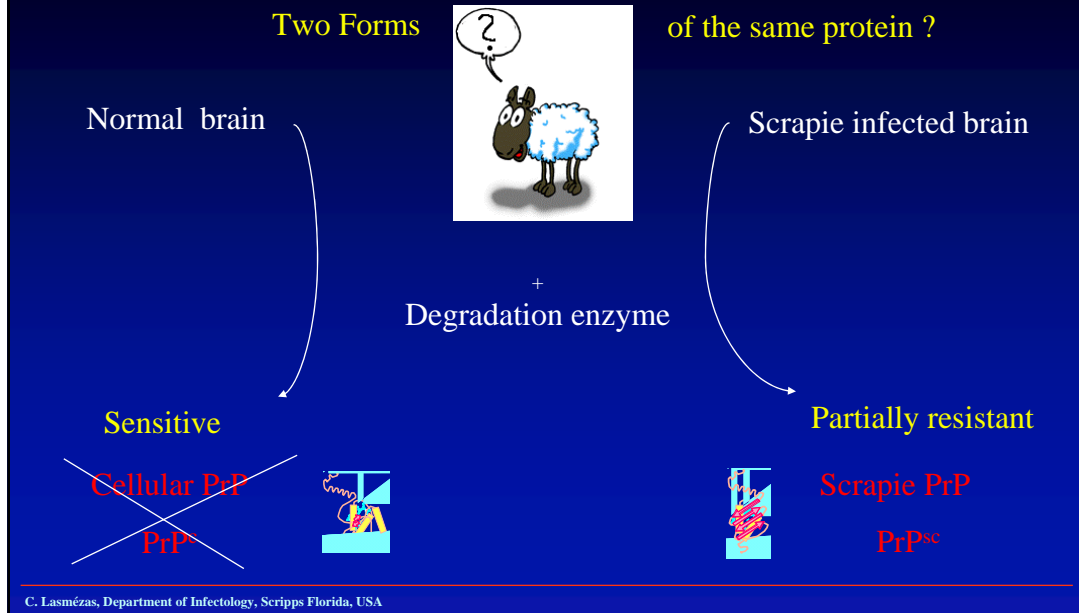
感受性

~~細胞 PrP~~
~~PrP^C~~



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How can a host protein be infectious ?



どのようにして宿主たん白質は感染可能となるのか？

