Prion

Definition:

prions are infectious particles encoded by gene of normal host cells , they are composed solely of protein;

they are implicated as the cause of certain “slow virus disease called transmissible spongiform encephalopathies(TSEs) disease” in animals and human.

The prion protein PrP exists in two different conformational forms

- **PrPc is thought to be the benign form of the protein and is found in normal, healthy cells.**

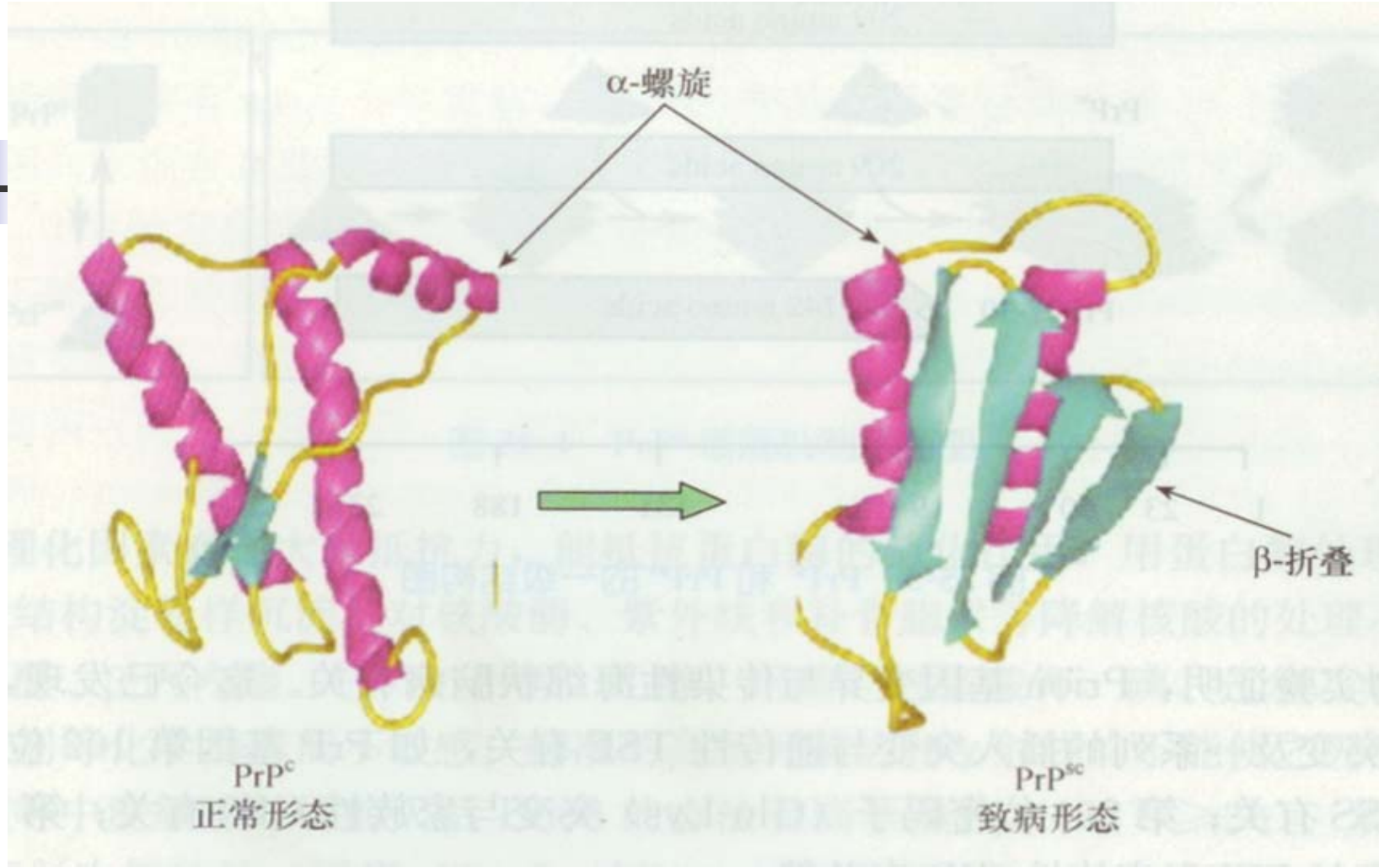
- **PrPsc is thought to be the infectious “scrapie” form which causes transmissible spongiform encephalopathies(TSEs) , such as bovine spongiform encephalopathy (BSE), Creutzfeldt-Jakob disease (CJD), and Kuru.**

PrPc (cellular PrP):

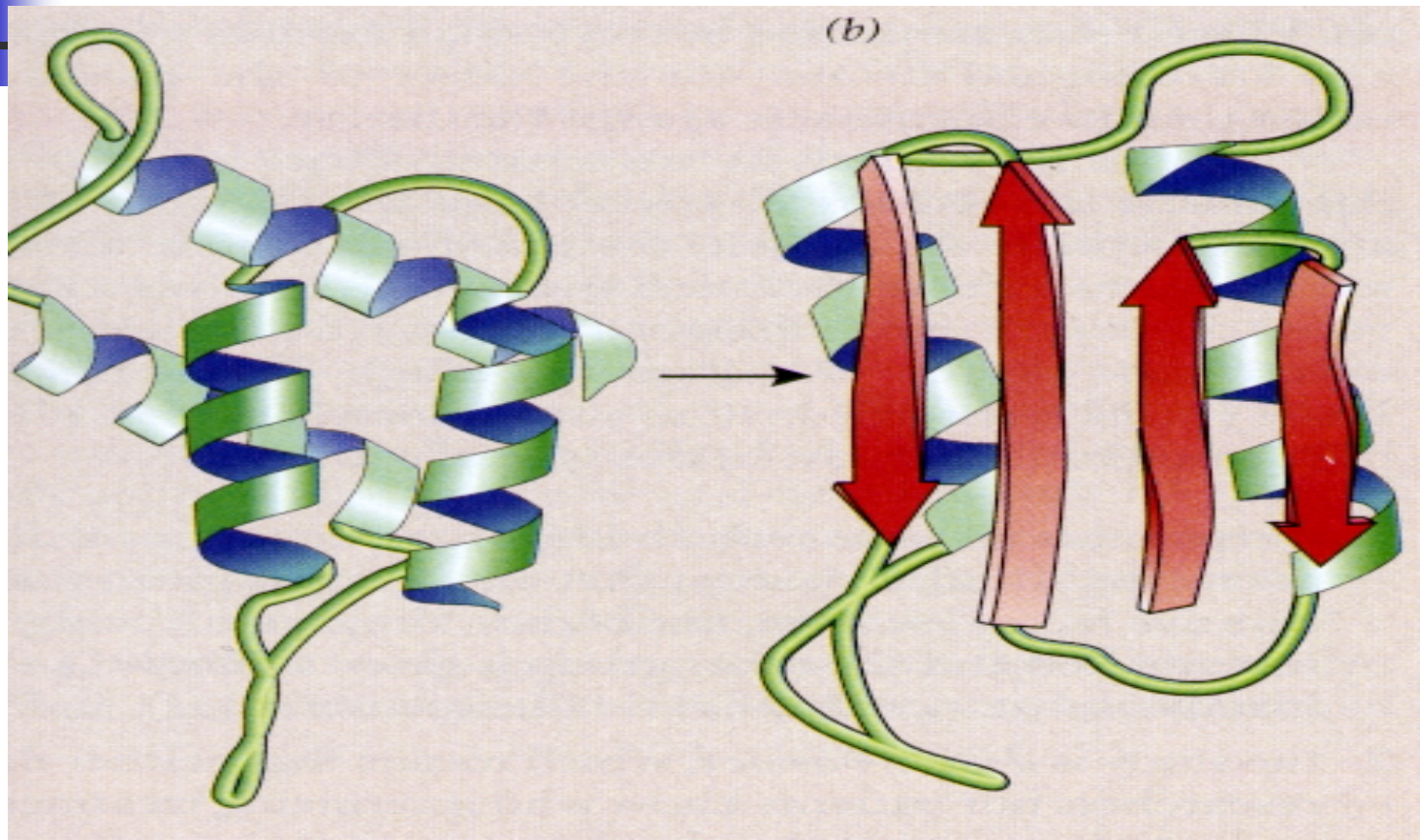
- The **normal** protein
- **Alpha helices**
- is **sensitive** to digestion by K protease

PrPsc (scrapie prion protein)

- The **disease-producing** protein
- **Beta conformation**
- is highly **resistant** to digestion by K protease



PrPC 和 PrPSC 的空间结构模式图



- PrP^{c}
- PrP^{sc}

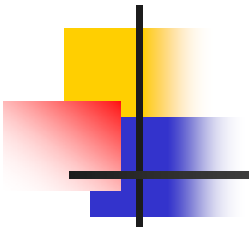
PrPc and PrPsc

	PrP c	PrP sc
soluble	+	–
K protease digesting	+	–
polypeptide chain	α-helical segment	ß-sheet
pathogenicity	**-**	**+**

- **Resistant:**

  strong,

  132 ℃ ≧ 2h



- **Pathogenesis & immunity**

- **Humans might be infected by prions in 2 ways:**
- **Acquired infection (diet and following medical procedures such as surgery, growth hormone injections, corneal transplants) i.e. infectious agent implicated.**
- **Apparent hereditary mendelian transmission where it is an autosomal and dominant trait. This is not prima facie consistent with an infectious agent.**

Prion Diseases

- Transmissible spongiform encephalopathies (TSEs) are a family of rare progressive neurodegenerative disorders that affect both humans and animals.
- They are distinguished by long incubation periods, characteristic spongiform changes associated with neuronal loss, and a failure to induce inflammatory response.

- **Human Prion Diseases**

*Creutzfeldt-Jakob disease (CJD)

* Variant Creutzfeldt-Jakob Disease (v-CJD)

*Gerstmann-Straussler-Scheinker Syndrome

*Fatal Familial Insomnia

*Kuru Disease

- **Animal Prion Diseases**

*Bovine spongiform encephalopathy (BSE)

*Scrapie of sheep and goat

*Chronic wasting diseaseof deer (CWD)

*Transmissible mink encephalopathy

*Feline spongiform encephalopathy

库鲁病（kuru disease）

疯牛病，与病牛接触或进食病牛肉是人v-CJD最主要的发病原因。