MANASH PRATIM DUTTA

Introduction

Background

Diseases

Types of Prions

Introduction

What is a Prion? (Pree-on)
Full Name: Proteinaceous infection particle

Definition

Features

Importance

Definition

Prion: a misfolded protein with the ability to independently reproduce and become infectious. They force other proteins to conform to there structure.

- Single molecules which contain 250 amino acids
- The human gene, PRNP, on chromosome 20 encodes for the prion proteins
- Prions have the ability to infect nearby normal proteins via contact
- extremely resistant to heat and chemicals
- They essentially act as viruses which lack nucleic material and are 100x smaller than the smallest known virus

- Prions are zoonotic and known for causing and spreading a variety of diseases in a wide range of organisms
- No current cure or antibiotics for any prion related diseases
- Prion related diseases are neurodegenerative and are always fatal
- It has broken parts of Darwin's evolutionary theory upon discovery
- In 2004-2007, a neurodegenrative disease known as Bovine Spongiform Encelphalopathy, or mad cow disease caused an epidemic in U.S. cattle costing the U.S. approximately 11 billion dollars due to inability to import beef.

Background

Discovery

Identification

Darwin's Theory

DISCOVERY

- Prion research began before prions were ever identified.
- Carelton Gajdusek studied a cannabalistic tribe in Papau New Guinea infected with Kuru, a type of neurodegenerative disease.
- At the time it was an unknown disease and would later become the first true discovery of a prion.

IDENTIFICATION

Stanley Prusiner

- A Researcher and neurologist at UC San Francisco.
- He discovered the major driving forces behind the identification and naming of the proin and there misfolded configurations
- This discovery alloted Dr. Prusiner the nobel peace prize

Darwin's Theory

- The research of Dr. Prusiner and similiar researchers disproved that life consists of the competition among individuals to spread there genetic information.
- This theory confused scientists as it was long believed that proteins were not "alive" and has no DNA to pass on.

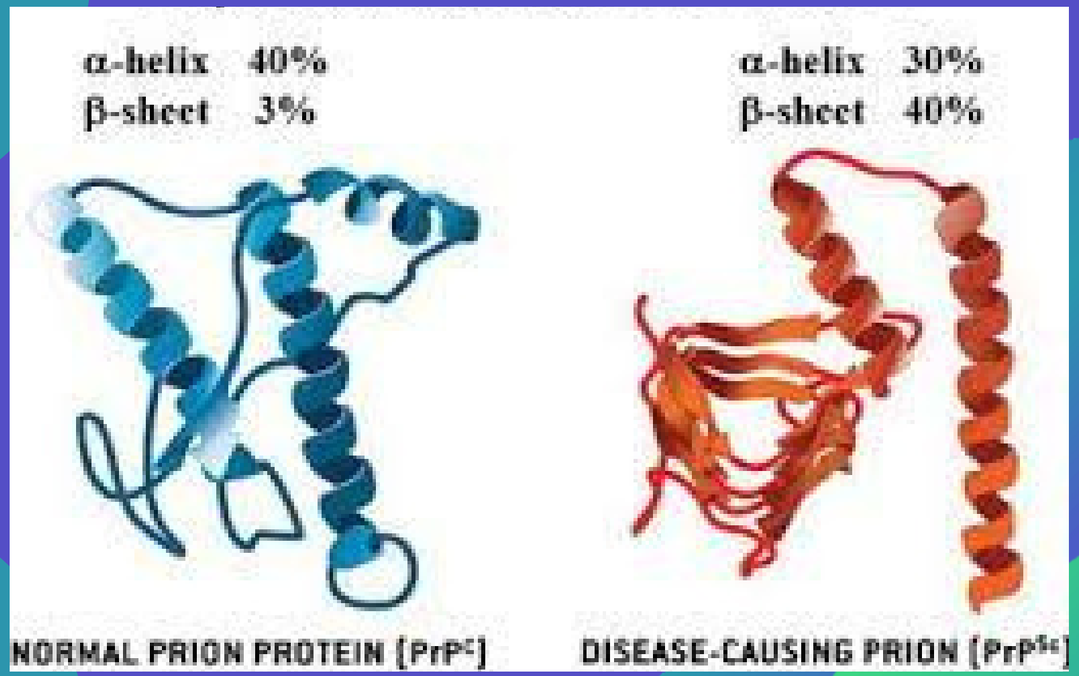
TYPES

Two types of prions:

PRP-sen: Normal Prion

PRP-res: Diseased Prion

PRP-sen



PRP-res

Also known as PRPc

- present on the surface of normal and healthy cells
- Found mainly in neurons and used during synapses
- Have been correlated with-------
  a) communication between neurons
  b) cell death
  c) sleep pattern controls

Sen stands for sensitive, these prions are sensitive to being broken down

Also known as PRPsc

- Form of prion which causes disease due to its misfolded configuration
- Causes organisms to develop Transmissible Spongiform Diesese's or TSE's such as mad cow disease, scrapies, and Kuru

Res stands for resistant and this type is resistant to being broken down by enzymes in the body

Allows for the accumulation of this type and eventual neuro-degerative disease

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Diseases

Prion Diseases

There are Four major prion-related diseases

1. Jacob Creutzfeld Disease:most commone prion disease in humans
2. Kuru: this is very rare human specific prion disease, found in a New Guinean tribe which is contracted by the consumption of infected human brain tissue.
3. Bovine Spongiform Encephalopathy (mad cow disease): most common prion disease in cattle
4. Chronic wasting Disease: most prion common disease in deer, elk, and moose.

THANK YOU