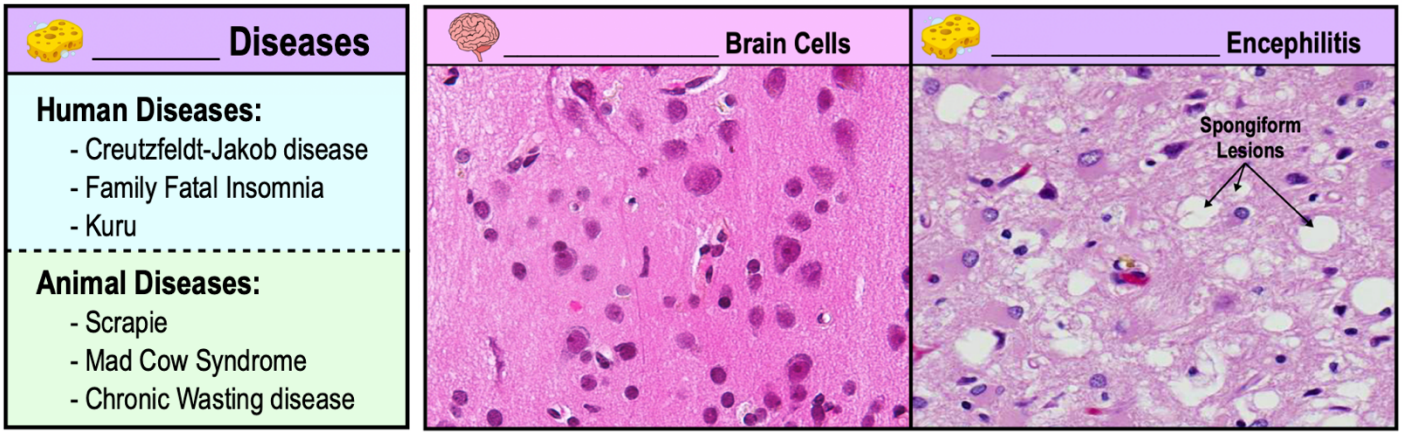
CONCEPT: PRIONS

●*Recall:* _____ (**Pr**oteinaceous **In**fectious Agents) *obligate intracellular parasites* made of only *misfolded* _____.

□ Cause the normal protein to *misfold* which can lead to a _____.

□ Prions accumulate in neural tissue (ex. _____) causing *transmissible spongiform encephalopathies.*

□ **Transmissible spongiform encephalopathies:** brain tissue deteriorates forming holes & a *sponge-like* appearance.



Prion Development in Neurons Leads to Scrapie

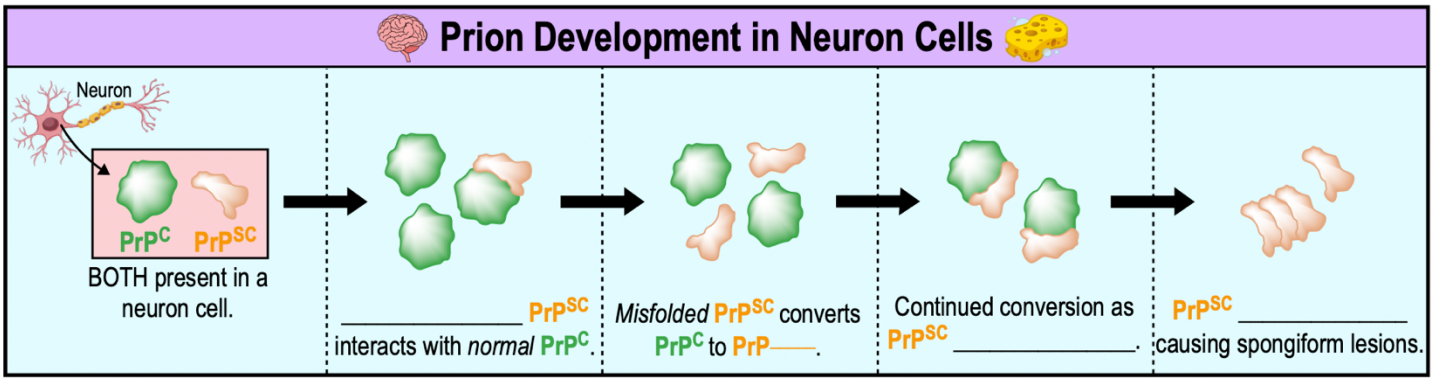
●The infectious prion protein is **PrP^{SC}** (**Pr**ion **P**rotein, **Sc**rapie) & normal its normal form is **PrP^{C}** (**Pr**ion **P**rotein, **C**ellular).

□ Accumulation of PrP^{SC} in the brain is attributed to its interaction with the normal protein _____.

□ The interaction causes the normal PrP^{C} protein to _____, resulting in a PrP^{SC} prion.

□ PrP^{SC} accumulates & begins to aggregate in the brain leading to spongiform encephalitis.

EXAMPLE: Prion development in neuron cells results in the neurological disease Scrapie in animals.



PRACTICE: What are prions?

a) Mobile segments of DNA.

b) Tiny circular molecules of RNA that can infect plants.

c) Viral DNA that attaches itself to the host genome and causes disease.

d) Misfolded versions of normal proteins that can cause disease.

CONCEPT: PRIONS

PRACTICE: Which of the following is true of prions?

a) They can be inactivated by boiling at 100 °C.

b) They contain a capsid.

c) They are a rogue form of protein, PrP.

d) They can be reliably inactivated by an autoclave.