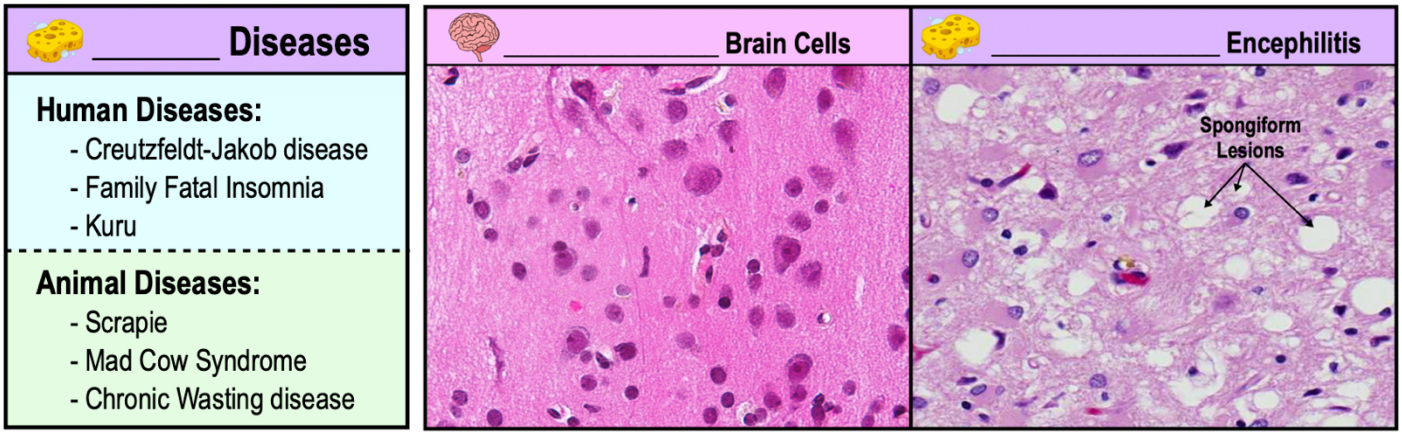
CONCEPT: PRIONS

●*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

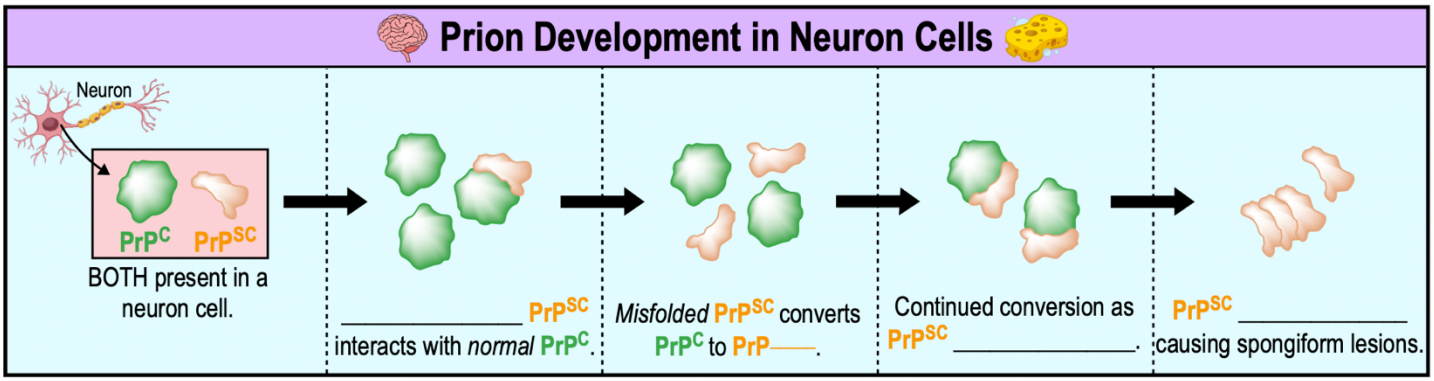
●The infectious prion protein is **PrP^{SC}** (**Pr**ion **P**rotein, **Sc**rapie) & normal its normal form is **PrP^{C}** (**Pr**ion **P**rotein, **C**ellular).

□ 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.

PRACTICE: Viroids and prions are both infectious agents. All of the following are major differences between viroids and prions except which of these answers?

a) Viroids are only known to infect plant cells while prions are known to infect plant and animal cells.
b) Viroids cause the misfolding of the host cell's proteins while prions block the host cell's expression of genes.
c) Viroids are nucleic acid molecules while prions are composed of polypeptide chains.
d) Viroids require the host's RNA polymerase to replicate while prions replicate by transforming the host cell's proteins.

PRACTICE: Mad cow disease is a prion disease found in cattle. Why are prion diseases in meat a big concern?

a) Prions cannot be destroyed by cooking meat.
b) Prions cannot be destroyed with irradiation of meat.
c) Prions are the most difficult infectious agent to deactivate or denature.
d) There are no cures for prion diseases.
e) All of the above.