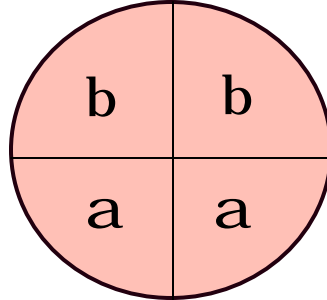


Sickle Cell Mutation

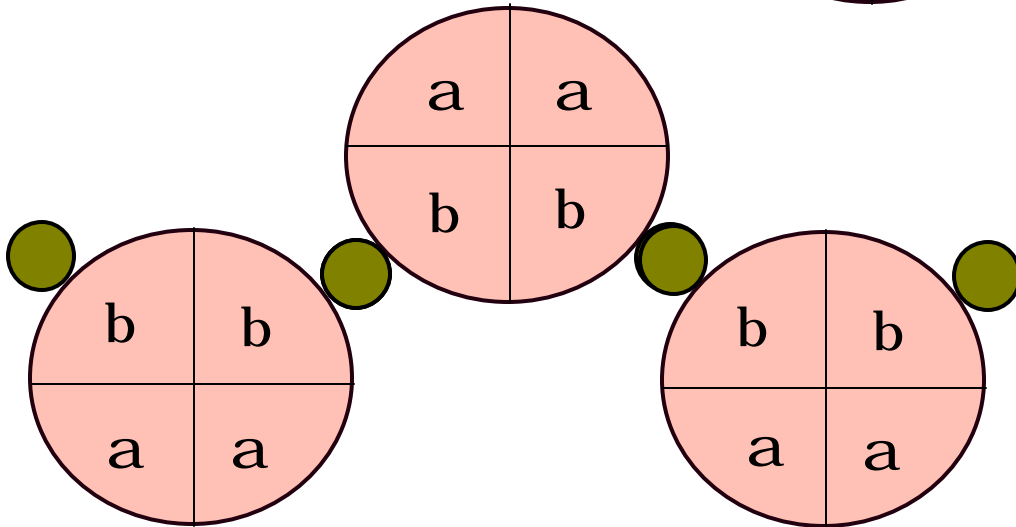
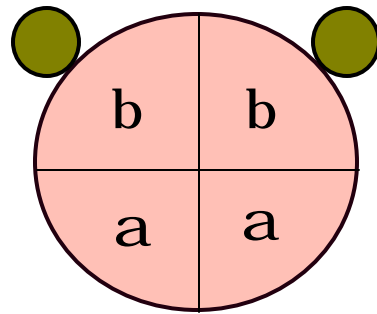
Hemoglobin is usually soluble in red blood cell



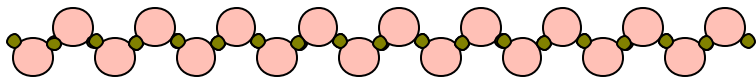
Sickle Cell Anemia

A **single** amino acid change

- **glu** to **val** on b subunits
- change causes non-polar patch
- adheres to other hemoglobin



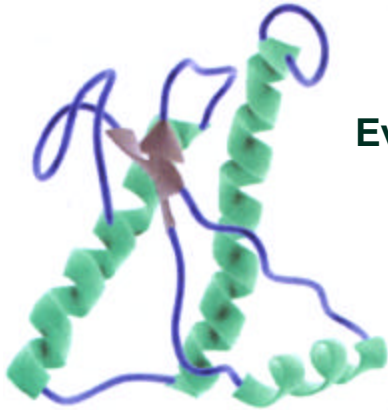
insoluble fibers of hemoglobin



Prions

Infectious proteins

- no DNA or RNA present
- causes Creutzfeldt-Jakob Disease
 - loss of memory
 - loss of motor skills
 - dementia
 - scraping and biting off of skin
 - invariably fatal



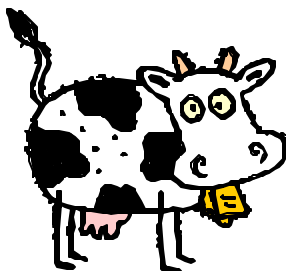
Everyone has normal protein in brain

- Prp
- mostly α -helical, soluble



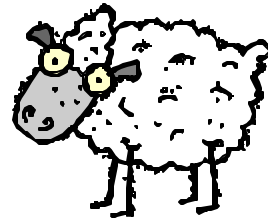
In patients with disease

- have Prp^{sc}
- same protein, different conformation
- mostly β -sheet, insoluble
- altered protein converts the normal protein to the altered conformation



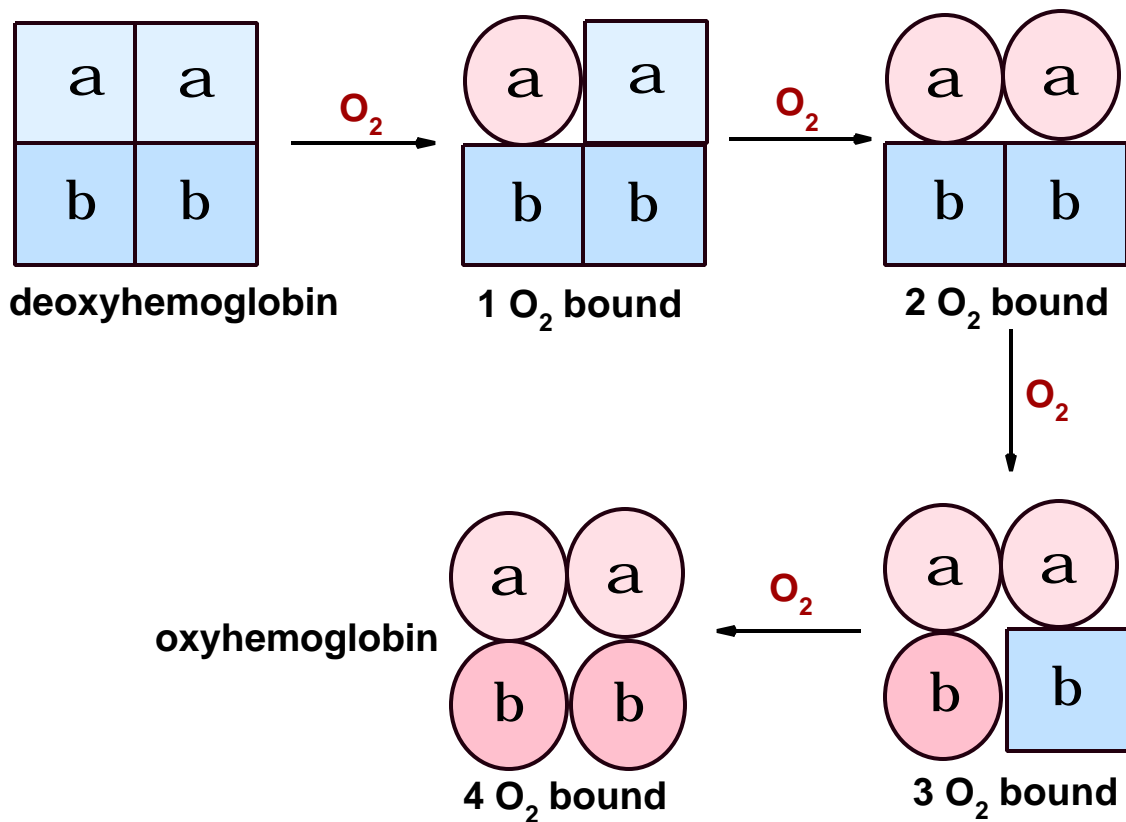
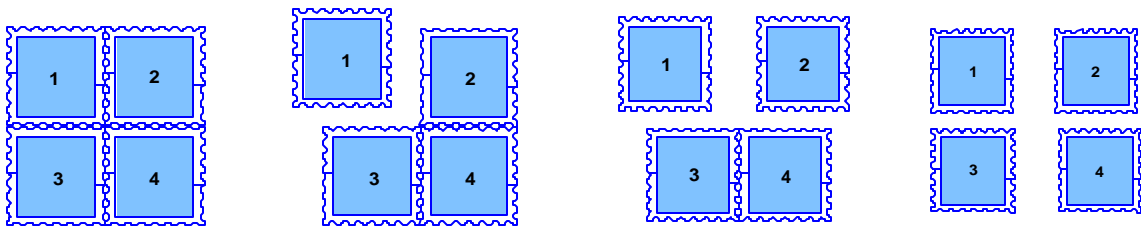
Catch this disease....

- hereditary
- sporadic
- eating infected meat (human or cow)



Hemoglobin (cooperativity)

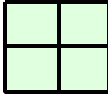
Each hemoglobin has 4 subunits, 2a and 2b
 Each hemoglobin can bind 4 molecules of O₂
 Binding of each O₂ makes binding the rest easier
 Called Cooperativity

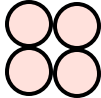


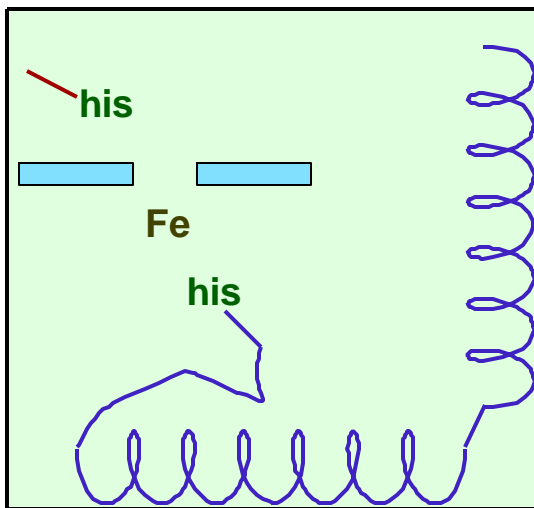
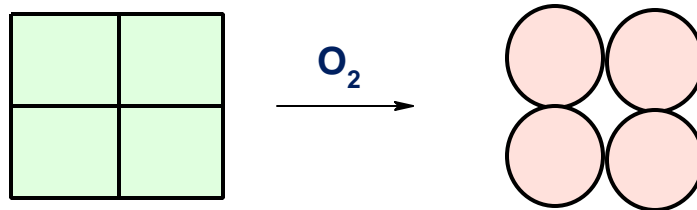
Hemoglobin Conformational Change



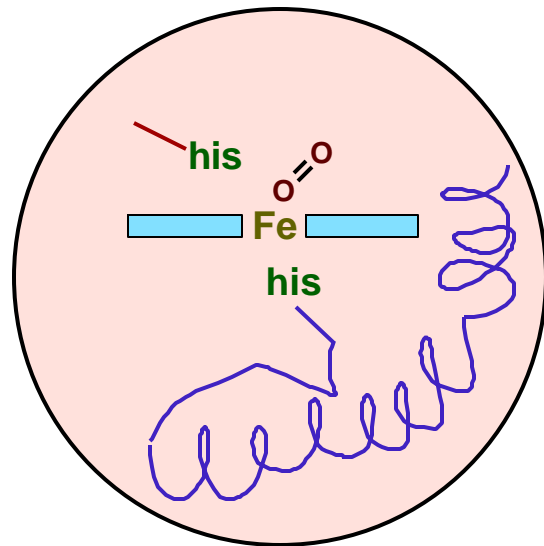
Change in 3° and 4° structure upon O₂ binding.

deoxyhemoglobin → T form (tense) 

oxyhemoglobin → R form (relaxed) 



deoxyhemoglobin



oxyhemoglobin