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A protein variant that is the cause of sickle cell anemia is a heterotetrameric protein. The protein undergoes a process of denaturation known as denaturation. We'll talk about why those things cause changes. and a process that a lot of people do n't necessarily think about but as engineers some of you will. The proteins stretch out into their denatured state. and instead of refolding to a compact structure, they just start aggregating with each other. Salts and organics may slip into a hydrophobic core and break them up. And then too much of a high concentration of an organic solvent that is miserable with water. A defect in a transport protein that carries oxygen around the body is a defect in hemoglobin. These diseases are what are known as inborn errors of metabolism. So these are tiny changes in the protein that cause dramatic changes in its structure and function of the protein. So hemoglobin causes a real problem with the quaternary structure and causes proteins to aggregate.

The iron heme complex is responsible for the red color of your blood cells. Hemoglobin, a tetrameric protein, has cooperative oxygen binding in a very narrow range of oxygen, which allows it to fill all four sites in the protein with an oxygen molecule. There are four proteins, beta globin, alpha globin, and beta globins, that are 146 amino acids long in each of them. A single defect in beta globine alters the properties of the hemoglobin and causes sickling of your red blood cells. One change in beta globin means two changes in the structure of the whole structure of hemoglobin. The mutation alters the way hemoglobin behaves. Sickle red blood cells have evolved to move smoothly through your capillaries. As soon as you get a different shape that's sort of not that discoid structure, they start clogging in the capillary. And when you have the defect where all of your hemoglobin is messed up with this variation, it is incredibly painful. People who are homozygous for the defect are often hospitalized with a lot of transfusions.

This is a dimer of tetramers. And if I just show you just some of the subunits, I can actually show you how there's two of each subunit in each structure. So what I can do is I can show you everything as a cartoon and get rid of all those little lines. And then you can see the structure where you see two beta globins and two alpha globins. Phenylalanine and leucine are both hydrophobic, providing a patch on the one tetramer where the valine from the other tetramer