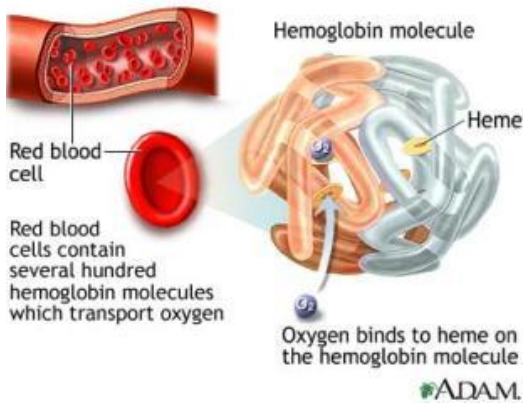
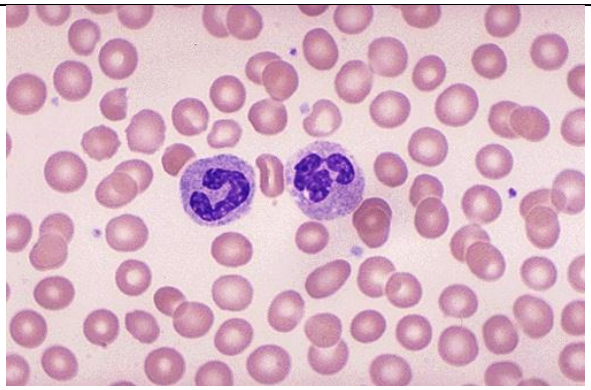
4.1 What is Sickle Cell? Study Guide by Hisrich

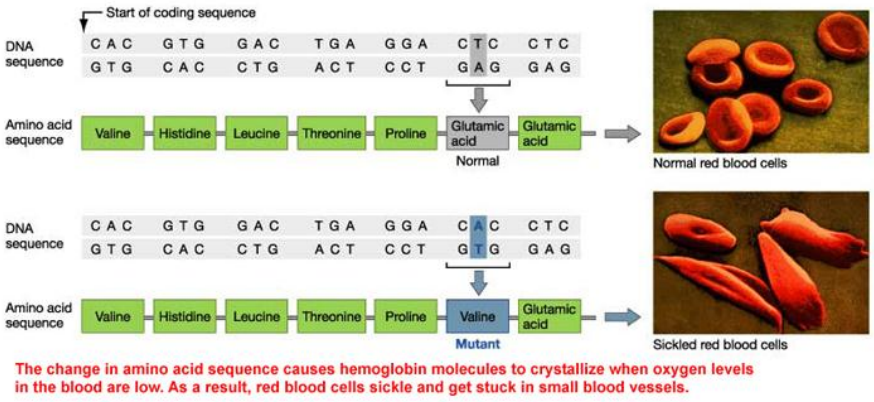
4.1.a. How do cells get the oxygen they need for energy production?

Erythrocytes (red blood cells) contain a **protein** called **hemoglobin** (“round blood”)—hundreds of molecules of it, actually. **Hemoglobin** binds to oxygen, picking it up from the alveoli and dropping it off in capillary beds throughout the bodies tissues. **Hemoglobin** is also the **protein** that picks up the carbon dioxide waste produced by all cells and brings it back to the alveoli so that the respiratory system can remove it from the body.	

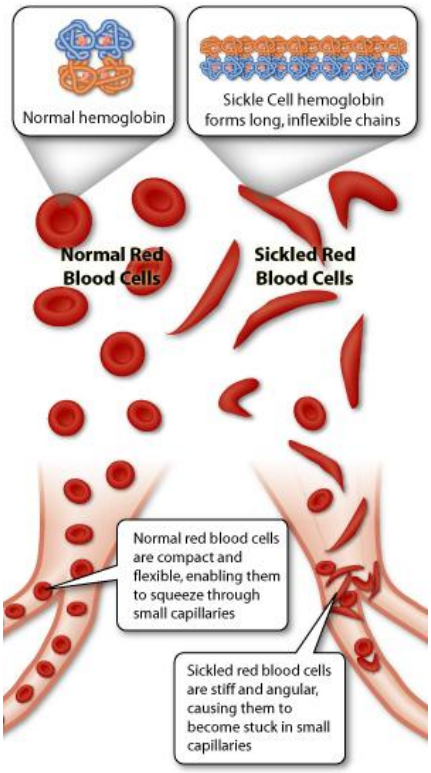
4.1.b. What do normal red blood cells look like when placed under a microscope?

	Normal red blood cells are round, but sort of flat in the middle (to increase surface area). The slide shown left is from a person with normal red blood cells. Most of the cells shown are erythrocytes. The large ones in the center are leukocytes (white blood cells) and the specks are thrombocytes (platelets).

4.1.c. Why do some people have differently shaped red blood cells?

People with **sickle cell anemia** have red blood cells with an abnormal shape. They are called “sickle” because they are shaped like the cutting tool called a sickle.		**Sickle cell anemia** *(“no blood”)* is a recessive genetic **trait** that must be inherited from both parents. It’s called anemia because the blood lacks normal **hemoglobin**. **Hemoglobin** contains two parts—**alpha globin** and **beta globin**. In a person with **sickle cell anemia**, there is a mutation in the **beta globin** portion, resulting in the substitution of ONE incorrect **amino acid**, causing the entire **protein** to fold incorrectly.

4.1.d. What effect does the altered shape of the red blood cell have on the health of the individual?

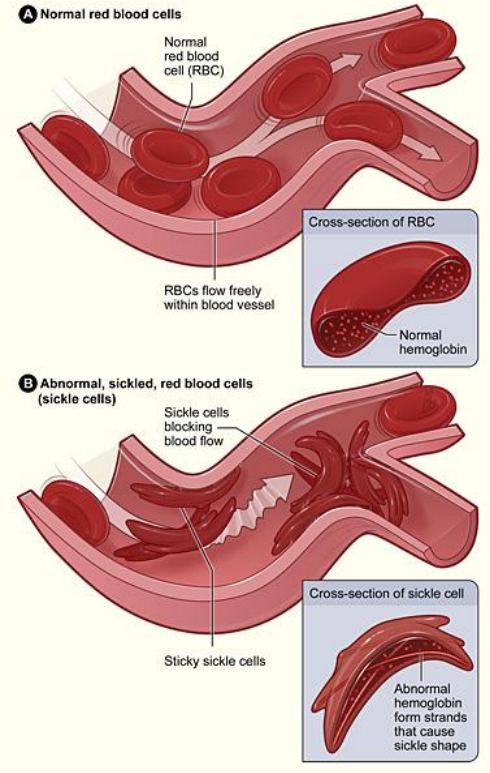


The **hemoglobin protein's** abnormal shape causes the red blood cells to have an abnormal shape. The can still carry oxygen, but they tend to get stuck in capillaries and make it difficult for blood to circulate to all the body's tissues.

The primary **symptom** of **sickle cell anemia** is pain, which is caused by lack of oxygen/nutrients to the body tissues.

Complications of **sickle cell anemia** include swelling of hands and feet, enlargement of the spleen, increased infections, acute chest syndrome (like pneumonia), eye problems & more.

The **prognosis** for a person with **sickle cell** is that there is no cure. Medications can treat the symptoms. Bone marrow and stem cell transplants can also reduce the effects.



4.1.e. What is the difference between someone having the **sickle cell trait** and having **sickle cell anemia**?

Sickle Cell Trait	Both	**Sickle Cell Anemia**
Normal **hemoglobin**	Red blood cells have **hemoglobin**	Abnormal **hemoglobin**
Normal red blood cells	Have red blood cells	Sickle shaped red blood cells
Inherited one copy of the mutation	Inherited the gene from parent(s)	Inherited 2 copies of the mutation
No ill effects	Have protection from malaria	Many health complications
	Occurs mostly in people descended from those in the tropics	

4.1.f. Where in the world does **sickle cell** disease occur most often?

Sickle cell anemia occurs most in the tropics, in places like South America & Central Africa. It occurs there because the mutation protects from malaria, and malaria is a big killer in the tropics (temperatures allow mosquitos to thrive). There is no survival benefit to the trait in more Northern climates, so the disease wouldn't persist here. However, due to immigration **sickle cell** disease is a problem even in places like the United States. It is a problem in people whose ancestors come from the affected regions (African Americans, Indian Americans, Middle Eastern immigrants, South American immigrants, etc). **Sickle cell** is virtually non-existent in people of European descent.