

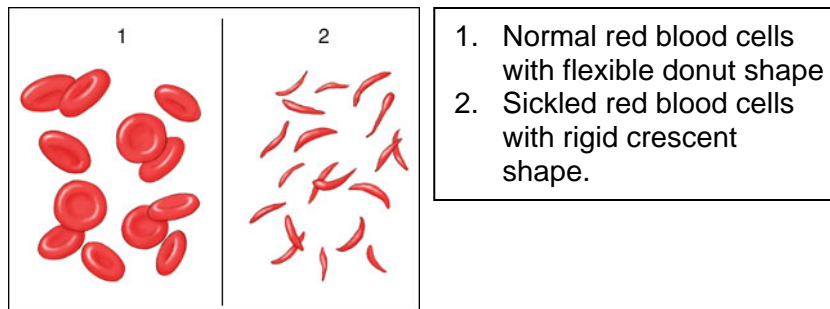
WHAT IS SICKLE CELL DISEASE?

Sickle cell disease is a common and often life threatening disease. It is an inherited, non-contagious premature destruction of red blood cells called hemolytic anemia. Hemoglobin is a complex molecule and the most important part of red blood cells. Sickle cell disease occurs from genetic abnormalities in hemoglobin:

- Normal red blood cells contain molecules called *hemoglobin A*
- In a sickle red blood cell, a variant of this molecule exists called *hemoglobin S* (S is for sickle)

The difference between hemoglobin A (HbA) and hemoglobin S (HbS) lies in only one protein out of about 300 that are common to both. However, this tiny abnormality can have a major impact. Sickle cell disease is a result of changes in hemoglobin S:

- The destructive nature of the sickle hemoglobin develops when it loses oxygen
- Under certain conditions (eg. acidosis, dehydration, infection and low oxygen, etc) the red blood cells form rigid rods that distort the red blood cell into a sickle or crescent shape
- These abnormally sickle shaped cells are both rigid and sticky. They cannot squeeze through capillaries and thus block the flow of blood, depriving tissues and organs of the oxygen they need
- Sickled red blood cells can become trapped within the blood vessel and interfere with normal blood flow. This obstruction can lead to sudden pain anywhere in the body as well as cause damage to body tissues and organs over time
- Dehydration can also result because excessive acidity and abnormal shape of the sickle cell causes water and potassium loss from the cell
- Sickle cells also have a shorter life span (10 to 20 days) than that of normal red blood cells (90 to 120 days)
- The body produces new red blood cells every day but sickle cells are destroyed so fast that the body cannot keep up. The result is a drop in red blood cell count called *anemia*



ADDITIONAL RESOURCES:

Sickle Cell Disease - http://sickle.bwh.harvard.edu/menu_sickle.html

The Sickle Cell Information Center - <http://www.scinfo.org/>

Sickle Cell Disease: An Overview - <http://www.mcg.edu/centers/sicklecell/overview/index.htm>

Department of State Health Services - Texas - <http://www.dshs.state.tx.us/newborn/sickle.shtm>