

Sickle cell disease is an inherited blood disorder that affects red blood cells and is not uncommon in the African-American community.

One of the questions that is on the current NCHSAA pre-participation medical form has to do with sickle cell disease, so it is important that everyone involved in high school athletics has some awareness and understanding about the disease.

What is Sickle Cell Disease?

People with sickle cell disease have red blood cells that contain mostly hemoglobin* S, an abnormal type of hemoglobin. Sometimes these red blood cells become sickle-shaped (crescent shaped) and have difficulty passing through small blood vessels.

When sickle-shaped cells block small blood vessels, less blood can reach that part of the body. Tissue that does not receive a normal blood flow eventually becomes damaged. This is what causes the complications of sickle cell disease. There is currently no universal cure for sickle cell disease.

Hemoglobin – is the main substance of the red blood cell. It helps red blood cells carry oxygen from the air in our lungs to all parts of the body. Normal red blood cells contain hemoglobin A. Hemoglobin S and hemoglobin C are abnormal types of hemoglobin. Normal red blood cells are soft and round and can squeeze through tiny blood tubes (vessels). Normally, red blood cells live for about 120 days before new ones replace them.

People with sickle cell conditions make a different form of hemoglobin A called hemoglobin S (S stands for sickle). Red blood cells containing mostly hemoglobin S do not live as long as normal red blood cells (normally about 120 days). They also become stiff, distorted in shape and have difficulty passing through the body's small blood vessels. When sickle-shaped cells block small blood vessels, less blood can reach that part of the body. Tissue that does not receive a normal blood flow eventually becomes damaged. This is what causes the complications of sickle cell disease.

Types of Sickle Cell Disease

There are several types of sickle cell disease. The most common are: Sickle Cell Anemia (SS), Sickle-Hemoglobin C Disease (SC), Sickle Beta-Plus Thalassemia and Sickle Beta-Zero Thalassemia.

What is Sickle Cell Trait?

Sickle Cell trait (AS) is an inherited condition in which both hemoglobin A and S are produced in the red blood cells, always more A than S. Sickle cell trait is not a type of sickle cell disease. People with sickle cell trait are generally healthy.

Inheritance

Sickle cell conditions are inherited from parents in much the same way as blood type, hair color and texture, eye color and other physical traits. The types of hemoglobin a person makes in the red blood cells depend upon what hemoglobin genes the person inherits from his or her parents. Like most genes, hemoglobin genes are inherited in two sets...one from each parent.

Examples:

If one parent has Sickle Cell Anemia and the other is Normal, all of the children will have sickle cell trait.

If one parent has Sickle Cell Anemia and the other has Sickle Cell Trait, there is a 50% chance (or 1 out of 2) of having a baby with either sickle cell disease or sickle cell trait with each pregnancy.

When both parents have Sickle Cell Trait, they have a 25% chance (1 of 4) of having a baby with sickle cell disease with each pregnancy.

Who Is Affected?

In the United States people are often surprised when they learn that a person who is not African American has sickle cell disease. The disease originated in at least 4 places in Africa and in the Indian/Saudi Arabian subcontinent. It exists in all countries of Africa and in areas where Africans have migrated.

It is most common in West and Central Africa where as many as 25% of the people have sickle cell trait and 1-2% of all babies are born with a form of the disease. In the United States with an estimated population of over 270 million, about 1,000 babies are born with sickle cell disease each year. In contrast, Nigeria, with an estimated 1997 population of 90 million, 45,000-90,000 babies with sickle cell disease are born each year.

The transatlantic slave trade was largely responsible for introducing the sickle cell gene into the Americas and the Caribbean. However, sickle cell disease had already spread from Africa to Southern Europe by the time of the slave trade, so it is present in Portuguese, Spaniards, French Corsicans, Sardinians, Sicilians, mainland Italians, Greeks, Turks and Cypriots. Sickle cell disease appears in most of the Near and Middle East countries including Lebanon, Israel, Saudi Arabia, Kuwait and Yemen.

The condition has also been reported in India and Sri Lanka. Sickle cell disease is an international health problem and truly a global challenge.

<http://www.sicklecelldisease.org/index.phtml>