

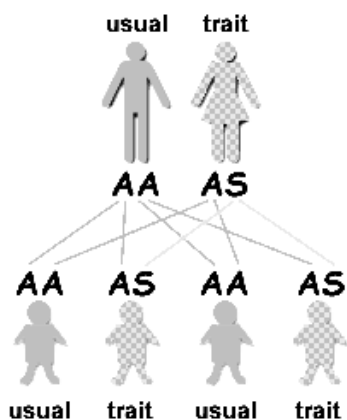
Sickle Cell: It's Your Choice

What Does "Sickle Cell" Mean?

Sickle is a type of hemoglobin. Hemoglobin is the substance that carries oxygen in the blood and gives blood its red color. A person's hemoglobin type is not the same thing as blood type. The type of hemoglobin we have is determined by genes that we inherit from our parents. The majority of individuals have only the "normal" type of hemoglobin (A). However, there are a variety of other hemoglobin types. Sickle hemoglobin (S) is one of these types.

There Are Two Forms of Sickle Cell.

Sickle cell occurs in two forms. Sickle cell trait is not a disease; Sickle cell anemia (or sickle cell disease) is a disease.



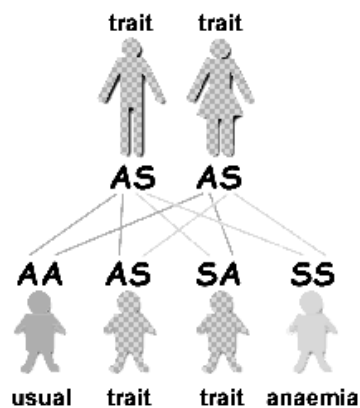
Sickle Cell Trait (or Sickle Trait)

Sickle cell trait is found primarily in African Americans, people from areas around the Mediterranean Sea, and from islands in the Caribbean. Sickle cell trait occurs when a person inherits one sickle cell gene from one parent and one normal hemoglobin gene from the other parent. A person with sickle cell trait is healthy and usually is not aware that he or she has the sickle cell gene. A person who has sickle trait can pass it on to their children. If one parent has sickle cell trait and the other parent has the normal type of hemoglobin, there is a 50% (1 in 2) chance with EACH pregnancy that the baby will be born with sickle cell trait.

When ONE parent has sickle cell trait, the child may inherit:

- 50% chance for two normal hemoglobin genes (normal hemoglobin- AA), OR
- 50% chance for one normal hemoglobin gene and one sickle cell gene (sickle cell trait- AS).

Sickle Cell Anemia (Sickle Cell Disease, or Hemoglobin SS)



Sickle cell anemia occurs when a person inherits two sickle cell genes, one from each parent. If both parents have sickle cell trait, there is a 25% (1 in 4) chance with EACH pregnancy that the baby will have sickle cell anemia. A child with sickle cell anemia appears normal at birth. However, later in infancy, anemia develops and the child tires easily. A second problem is attacks of pain called "crises". These crises can occur without warning, affect any part of the body, and last hours or days. A third problem is frequent infections and unexplained fevers. Daily penicillin and folic acid are required. Crises and infections may require hospitalizations and additional medications and may lead to significant numbers of absences from school or work. Over time damage to the organs (heart, kidneys, etc.) may occur. Sickle cell anemia does not cause mental retardation, but is a significant chronic health condition.

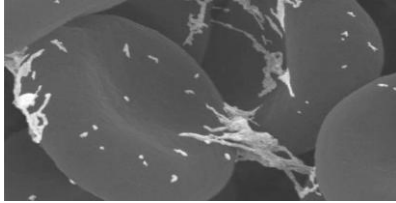
When BOTH parents have sickle cell trait, the child may inherit:

- 25% chance for two normal hemoglobin genes (normal hemoglobin- AA), OR
- 50% chance for one normal hemoglobin gene and one sickle cell gene (sickle cell trait- AS), OR
- 25% chance for two sickle cell genes (sickle cell anemia- SS)

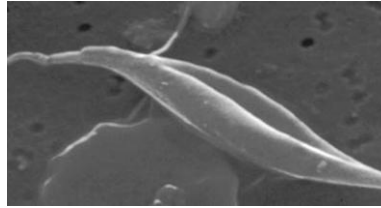
Why is it Called “Sickle Cell” Anemia?

In sickle cell anemia, the red blood cells have an abnormal appearance:

NORMAL



SICKLE CELL ANEMIA



This abnormal “sickle” shape causes the blood cells to sometimes get “stuck” in the arteries and veins of a person with sickle cell anemia, causing the painful crises and other medical problems.

There Are Other Variations of Sickle Cell.

Other types of sickle cell diseases are “sickle-hemoglobin C disease” and “sickle-beta-thalassemia”. Individuals with these conditions have inherited a sickle cell gene from one parent and a gene for either hemoglobin C or beta-thalassemia from the other parent. Symptoms of these forms may be similar to the symptoms for individuals with Hemoglobin SS Disease.

Who is Most Likely to Have Sickle Trait?

Sickle hemoglobin is most often found in people whose ancestors come from Africa, the Arabian States, South India and from countries around the Caribbean Sea, such as Puerto Rico, Cuba, Haiti and Jamaica, and also from countries around the Mediterranean Sea, such as Italy, Cyprus, Greece, Turkey, and Syria. Sickle cell trait occurs in approximately 1 in 12 (8%) African-Americans. Sickle cell disease occurs in approximately 1 in 375 African-Americans and affects more than 50,000 Americans.

How Can I Find Out If I Have Sickle Cell Trait?

There is a simple and accurate test for sickle cell trait. It requires a sample of blood taken from a vein. However, this test may not be done routinely and should be requested. The test is called hemoglobin electrophoresis.

What Can I Do If I Have Sickle Cell Trait?

If you have sickle cell trait, you need not be concerned for your own health. However, you may be at risk for having a child with sickle cell anemia. If your partner is tested and has normal hemoglobin test results, then your children would not be at risk to have sickle cell disease. But if your partner also has sickle cell trait, each of your children has a 25% (1 in 4) chance to inherit the sickle cell gene from both of you and have sickle cell disease.

If you have sickle trait, you should ask your partner to have hemoglobin electrophoresis. If you find out that you both have sickle cell trait during the first half of a pregnancy, you can consider whether you would like to have prenatal testing to see if the baby will have sickle cell anemia. Prenatal testing is optional. It is available for women who would want to know during the pregnancy if the baby will have sickle cell disease. All babies are tested right after birth to see if they have sickle cell anemia as part of their “Newborn Screening” test. Your physician can discuss the different testing options further with you, and may refer you to see a genetic counselor if you and your partner both have sickle trait.

For additional information, call Reproductive Genetics at 585-487-3480.

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