How common is sickle cell anemia? [1]

Dear Alice,

What percentage of African Americans in the U.S. are diagnosed with sickle cell anemia?

Answer

Dear Reader,

Although sickle cell anemia is a relatively rare inherited condition, its significant impact on the African American community and its life-threatening consequences have made it one of the better-known genetic disorders. About 70,000 to 100,000 people in the U.S. have sickle cell anemia. Of those affected, about 1 in 500 are African American. Relatedly, among the two million Americans that have the sickle cell trait (SCT), 1 in 12 of those are African American which means they don’t have the disease, but they can pass it on to their children. Even though both sickle cell anemia and trait are most common among African Americans, other Americans who trace their ancestry back to the Mediterranean region, South and Central America, Saudi Arabia, and India can also be affected.

Sickle cell anemia (one of the most common types of sickle cell disease) is caused by inheriting a defective hemoglobin gene from both parents. Hemoglobin is the part of red blood cells that is responsible for carrying oxygen in the blood stream throughout the body. Defective hemoglobin produced by the sickle gene become stiff, causing the red blood cells to change from rounded, doughnut-shaped cells that can flow easily through small blood vessels, to pointy crescent or sickle-shaped cells. They also have a shorter lifespan of 10 to 20 days rather than the typical red blood cell, which dies after about 120 days. Because the sickle-shaped cells die so quickly, the body can’t keep up with the production. The anemia that results (a condition caused by a subnormal level of hemoglobin or red blood cells) often leads to tiredness, jaundice, irritability, pale skin and may also result in delayed puberty and slowed growth for children.

Additionally, pain is also common for those with sickle cell anemia. When sickle-shaped cells form and create roadblocks in the blood vessels, a person has what is called a sickle cell crisis (also known as a painful crisis). Sickle cell crises often come on suddenly and can last from a few hours to a week or more. The amount of pain caused by a crisis can be mild and controlled by over-the-counter pain medications, or can be so excruciating that hospitalization and IV pain medications are required. Repeated crises have the potential to cause organ damage and can lead to a number of conditions and complications. However, adopting healthy lifestyle habits including a nutritious diet, regular (but not intense) exercise, adequate sleep, staying hydrated, avoiding circumstances that are too hot or too cold, getting your flu
shot and other vaccines can all help reduce the frequency and severity of sickle cell crises. Chronic pain in the bones is also common, lasting for weeks or months which can significantly hinder daily functioning.

Current treatment of sickle cell anemia consists of antibiotic regimens to fight infection, pain management strategies, and blood transfusions, when necessary. Newer, promising treatments include blood and bone marrow stem cell transplants, gene therapy, and various new medications. Research is ongoing to further investigate other treatment options. Every state in the U.S. now routinely screens newborns for sickle cell disease. Early detection and treatment of sickle cell anemia are, in part, responsible for increasing the life expectancy of those who have sickle cell anemia well into their forties, fifties, and beyond.

For more information about the condition, check out the Sickle Cell Disease Association of America [2] website.

Alice!
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