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The Burden of Sickle Cell Disease for Patients and Their Families

Announcer:
Welcome to CME on ReachMD. This episode is part of our MinuteCME curriculum.

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Dr. Andemariam:
This is CME on ReachMD, and I'm Dr. Biree Andemariam. In this episode, I will be discussing the burden of sickle cell disease for patients and their families.

So I think it's important to understand a little bit about the epidemiology of sickle cell disease, and know that it's the most common inherited blood disorder worldwide. It's inherited by simple Mendelian inheritance. It's autosomal recessive. So if 2 parents carry hemoglobin traits, such as sickle cell trait, they have a 1 in 4 chance with every birth of having a baby with sickle cell disease. Sickle cell disease actually affects millions of people worldwide. There's an estimated 300,000 births per year, and it's believed that by the year 2050, that this will increase to 400,000 births per year. In the U.S., the disease prevalence is estimated to be about 100,000 Americans living with sickle cell disease. And the mean survival really hasn't changed in a long time. The mean survival is still into the 40s. One in 365 African American babies is born with sickle cell disease. Sickle cell trait is also very common, it affects 1 in 100 Latinos, 1 in 12 African Americans, and even in my own state of Connecticut, newborn screening reveals that 1 in 60 births gives rise to a baby with sickle cell trait.

So what is sickle cell disease? Well, in order to understand that, you have to understand hemoglobin. Hemoglobin is made up of 2 alpha chains and 2 beta chains. And it's the beta chain that is problematic in sickle cell disease because people inherit a simple point mutation in their beta-globin gene. This leads to sickling of red blood cells under deoxygenated conditions. And these sickled red blood cells begin to occlude the blood vessels as they circulate through the body. This leads to hemolysis, worsening anemia, as well as nitric oxide depletion, inflammation, and resultant vasculopathy. There are several forms, including the most common form which is hemoglobin SS.

There are a lot of clinical consequences of having sickle cell disease. A lot of these are subclinical, meaning that they never really come to medical attention until there's a big problem. And this is because sickle cell disease affects virtually every organ in the body, and it can lead to chronic organ damage over time. There are also acute problems related to vaso-occlusion which can give rise to significant morbidity. These include vaso-occlusive crises such as pain crises, and the acute chest syndrome, and priapism, as well as acute stroke. The early mortality associated with sickle cell disease is largely due to infection, the acute chest syndrome, and even sudden death.

Sickle cell disease is also unpredictable in terms of its acute complications, as well as the accumulation of chronic complications that can lead to social, emotional, and sometimes even financial burdens for both the individual living with sickle cell disease, as well as his or her family and caregivers. As an example, for children, this often leads to days missed from school. For adults, this leads to days missed from either higher education or work. And for all of those people living with sickle cell disease, it makes it really difficult to make plans because they never know when an acute complication such as an acute painful episode is going to occur. This is an emotional drain. It's a financial drain. And research has demonstrated that living with sickle cell disease confers a worse quality of life than being

on dialysis.

This has been an introduction to sickle cell disease with a special focus on its burden to both patient and caregivers. Unfortunately, our time is up. Thanks for listening.

Announcer:
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