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ReachMD
www.reachmd.com
info@reachmd.com
(866) 423-7849

Making the Transition: Managing Patients with Sickle Cell Disease Across the Lifespan

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 lifelong management of patients with sickle cell disease, focusing on issues such as fertility and childbearing, as well as discussion with patients and caregivers and families as complications arise, or as new therapies emerge.

So it's important to note that there are many issues related to the lifelong management of people living with sickle cell disease, and I'd like to discuss some of them.

So first is fertility. I think it's important that people know that the data suggests that men and women with sickle cell disease actually have higher baseline rates of infertility. Some of this is due to their underlying disease, which might predispose men to hypogonadism as well as erectile dysfunction from repeated episodes of priapism. And in women, there's accelerated decline in ovarian reserve that we are just beginning to understand. There's also transfusion-related iron overload that can lead to gonadal failure in both genders.

What about childbearing? So in women, pregnancy actually is considered, by definition, a high-risk pregnancy. There are higher rates of adverse maternal as well as fetal outcomes that people really need to be knowledgeable about. These adverse outcomes include preeclampsia, preterm birth, intrauterine growth retardation, hemolytic disease of the newborn, and increased rates of neonatal ICU stays. All women with sickle cell disease who are pregnant really should be comanaged by a hematologist as well as a high-risk maternal fetal medicine specialist. This will maximize good outcomes of both mother and baby. And this should begin in adolescence. It's really important that they understand the inheritance of sickle cell disease, and how they might be able to reduce the risk of having children of their own with sickle cell disease.

It's really also very important for all of us to know that there are emerging complications that begin in adulthood. And with age, what we see is that there are increased rates of nephropathy, retinopathy, acute chest syndrome, bony infarcts, hemorrhagic stroke, pulmonary hypertension, cardiomyopathy, you name it. So a thorough review of systems with every visit can begin to identify some of the symptoms that will direct the testing that can be aimed at identifying organ involvement early. It's all about preserving organ function.

It's also important to note that as individuals with sickle cell disease live longer, we have to ensure that they undergo proper age-appropriate cancer and cardiovascular screenings.

Depression, anxiety, posttraumatic stress disorder, all of these mental health conditions are very common in individuals with sickle cell disease, and their incidence increases with age. We have to enlist the support of a social worker with expertise in mental health counseling, as well as psychological and psychiatric experts.

And as the treatment landscape evolves in sickle cell disease, it's critical to ensure that patients are aware of these new treatment options. Shared decision-making will optimize their adherence. And as such, efficacy of new treatments with a goal of making sickle cell

disease a chronic disease that can be managed is important.

It's also important that we keep in mind that most adults with sickle cell disease were actually never expecting to live past childhood. And the realities of this unanticipated survival are far-reaching and have direct impact on their reduced educational and professional achievement.

Also, the episodic disability related to intermittent pain crises leads to high degree of missed days from school and from work. And this leads to a significant pervasiveness of poverty among individuals living with sickle cell disease. It's really important to keep this in mind when caring for individuals with sickle cell disease.

Competing social issues can often make adherence to medical visits and treatments that much more challenging. It's important to enlist social work support to help identify potential social challenges that may improve adherence, communication, and clinical outcomes if identified.

I hope you have enjoyed this presentation, and that it provides you with insight into the lifelong issues that are faced by patients with sickle cell disease. Unfortunately, our time is up. Thanks for listening.

Announcer:
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