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Preventative Management of Sickle Cell Disease Complications

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

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Dr. Freiermuth:
This is CME on ReachMD. And I'm Dr. Caroline Freiermuth. Here with me today is Dr. Biree Andemariam. We will be addressing the preventative management of sickle cell disease complications.

Dr. Andemariam, how do you actively undertake preventative management of sickle cell disease complications in your clinical practice?

Dr. Andemariam:
So I think what's important to know that, is that close longitudinal follow-up with patients is really key. We have to ensure that patients are on appropriate disease-modifying therapy, starting with hydroxyurea as the backbone, and then adding on additional agents depending on whether anemia or vaso-occlusive crises are the defining phenotype in the individual patient.

Putting patients at risk for a first or subsequent stroke on chronic transfusional therapy is really important. It's also essential to monitor patients for iron overload and treat them if you find it, because untreated iron overload can have organ damage as a consequence, including the liver, as well as the heart. It's important to promptly assess and treat any patient presenting with a vaso-occlusive pain crisis and monitor them closely, because they're at risk for acute chest syndrome, which can be life threatening.

It's also important that we ensure that patients are seen by a primary care physician, and if the patient is female, to make sure that they're seeing an OB/GYN doctor for routine health maintenance. We also have to make sure that our patients are being referred to ophthalmologists to be seen at least once per year to screen them for sickle retinopathy, which can be effectively managed if caught early and not lead to blindness.

All patients with sickle cell disease should ideally be cared for by a multidisciplinary team. This is what ensures that all their medical and psychosocial issues are addressed by experts. The team should include hematologists as well as nurses who can address the sickle cell-specific issues as well as the social workers, psychologists, and even psychiatrists sometimes who can assess and treat comorbid mental health issues that actually are quite prevalent in this population.

It's also really critical that when pediatric patients age out of pediatric care, that there's a smooth transition process that begins during adolescence, and then culminates in the final transfer of care to an adult-focused sickle cell disease expert. Ensuring that every patient with sickle cell disease is cared for by a multidisciplinary team and then transitions properly from pediatric to adult care helps to reduce the barriers that can then optimize all the prevention efforts that we make as clinicians.

Dr. Freiermuth:
Thank you, Dr. Andemariam.

Emergency medicine focuses more on the acute management of disease rather than prevention. What we do need to be aware of is that

many complications related to sickle cell disease develop during a vaso-occlusive episode. Even if we have ruled things out at the start of the visit, if a patient's condition changes, we really need to start over and think, "What else did we miss? Is this the progression of the disease process? Is there some other complication that is now starting to develop?" Rerun our differential diagnosis, think about do we need to run further tests, do we need to do further imaging, so that we can really get to the bottom of what might be going on and not miss something important, that could have devastating effects for the patient.

We also know that recurrent presentations to the emergency department or to the hospital are common. So once we discharge a patient, we need to be really careful to give them good return precautions. We don't want someone to sit at home in pain and develop complications and think, "Well, they told me I was fine to go, so there's no point going back to the emergency department."

We also should ask questions about some of their social determinants of health. Do they have access to these outpatient medications that they've been prescribed? Do they have the ability to see a hematologist? Do they have a primary care physician? And so we really need to be checking in on some of those things.

Dr. Andemariam:
I 100% agree with you. And I think you as a practicing emergency medicine specialist oftentimes are the only one in the healthcare realm that are actually in contact with some of these patients with sickle cell disease.

Dr. Freiermuth:
Well, thank you. Hopefully, we get to the point where hospitals across the country really are opening those lines of communication between the emergency department and the outpatient world.

In this episode, we have discussed the preventative management of complications associated with sickle cell disease. I would like to thank Dr. Biree Andemariam for joining me today. Unfortunately, our time is up. Thanks for listening.

Announcer:
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