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A Quick Look at Standard Treatments for Sickle Cell Disease

Announcer:
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Dr. Andemariam:
This is CME on ReachMD, and I'm Dr. Biree Andemariam. Here with me today is Dr. Caroline Freiermuth. In this episode, we will be touching upon standard approaches to managing sickle cell disease. Dr. Freiermuth, could you please start the discussion addressing the management goals typically associated with sickle cell disease?

Dr. Freiermuth:
Sure, so I'm an emergency physician. And we're always taught to think worse first. Really, we want to be on high alert for any life-threatening complications. After we can quickly rule those out, then we move on to dealing with the presenting complaint. And so, you know, figuring out why the patient's there and what we can do to make their day better.

In the case for sickle cell disease, the most common complaint is vaso-occlusive episodes, and the most common reason they come in is for pain. And so we really want to make sure that we treat that pain quickly, and we treat it appropriately. In the emergency department, we must remember that many of these patients take opioids on a regular basis at home. And so sometimes we may need to adjust our dosing of opioids in the emergency department, taking that into account.

Acute chest syndrome causes a lot of morbidity and mortality for sickle cell disease. And so we really want to make sure that that is a diagnosis we don't miss. And finally, in kids, we really need to remember that patients with sickle cell disease are functionally asplenic, and we really need to think about the different bacteria that they may be infected with and tailor our antibiotics to that.

And what about your standard treatment in the outpatient world?

Dr. Andemariam:
Thank you, Dr. Freiermuth. Next, I'll provide a brief overview of the standard treatment approaches for sickle cell disease. These include, very importantly, some prophylactic strategies. So we're very fortunate in the United States that all 50 of our states test every newborn baby for sickle cell disease, and once they're identified, they're immediately put into care with an expert pediatric hematologist. One of the first things that's done is they're put on daily penicillin prophylaxis as well as they're ensured to have their pneumococcal vaccination and that's to make sure that they don't die from pneumococcal sepsis because they are particularly prone to that.

Children from the ages of 2 years to 16 years go through annual transcranial Doppler screening tests to look to see whether or not they're at risk of having a stroke. And if they are, they're put on a chronic monthly transfusional therapy to prevent that stroke from occurring. We also advocate that all people, children and adults alike, with sickle cell disease have routine ophthalmologic screening for sickle retinopathy to prevent the onset of blindness related to that retinopathy.

We also have four disease-modifying therapies that we can prescribe to individuals with sickle cell disease. Hydroxyurea has been around the longest. It's taken daily, and it reduces vaso-occlusive pain crises in people who take it. It works by increasing levels of fetal

hemoglobin which protects the red blood cells from sickling. In 2017, L-glutamine was approved, and that's a drug that can also reduce the frequency of vaso-occlusive pain episodes, and was also demonstrated to reduce incidence of the acute chest syndrome. In 2019, there were two drugs approved for sickle cell disease all in the same month. The first was crizanlizumab, it's a monoclonal antibody against P-selectin that's given intravenously approximately once per month. And it was approved on the basis of its ability to reduce the frequency of vaso-occlusive pain crises. Voxelotor is an oral anti-hemolytic agent, which is taken once per day. It works by binding to alpha-globin and keeping the hemoglobin within the red blood cells from being able to polymerize and cause sickling, anemia, and hemolysis.

We also use transfusions quite a bit in sickle cell disease. We use them both acutely for patients who present with either stroke or the acute chest syndrome. And we also use them chronically, giving patients transfusions every 2 to 4 weeks in the outpatient setting.

We also spend a lot of time managing both acute and chronic pain, which are the hallmark of the disease. And lastly, primary preventative care is so important.

This has been an exciting discussion on standard treatment approaches and management goals for sickle cell disease. I would like to thank Dr. Caroline Freiermuth for joining me today. Unfortunately, our time is up. Thank you for listening.

Announcer:
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