

**Minnesota Now (MPR) | Minnesota Now Minnesota Now -September sickle cell awareness month  
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TIM NELSON: Did you know that September is sickle cell awareness month? Sickle cell is a genetic trait affecting red blood cells, and it causes intense pain and anemia. Sickle cell disease occurs among one out of every 365 African American births, but some health care providers miss sickle cell symptoms or don't take seriously people who have the condition.

To learn more about why, I'm talking with Rae Blaylark and Dr. Stephen Nelson. Rae Blaylark is the President and CEO of the Sickle Cell Foundation of Minnesota, and Dr. Stephen Nelson is Chief Medical Officer at the foundation. Rae and Stephen, welcome to *Minnesota Now*.

**RAE BLAYLARK:** Thank you for having us.

**STEPHEN** Yes, thank you for having us.

**NELSON:**

**TIM NELSON:** So, I'd like to start with you, Rae. Why doesn't sickle cell disease get more attention?

**RAE BLAYLARK:** You know, I think, sickle cell disease doesn't get the attention that it deserves, because it's not talked about openly. It is-- there's still a stigma attached to it, it is still misunderstood as a Black disease, rather than a blood disease, and so, I think, that both on the health care side and on the community side, we need to be having more conversations like this.

**TIM NELSON:** And Dr. Nelson, let's get into a little more of the detail here. What does sickle cell actually do to the body and how dangerous is it?

**STEPHEN** Yeah. So, as you said, it's an inherited disorder, and actually 1 in 12 African American in the US is a carrier. They  
**NELSON:** don't have the disease, they're a carrier. It's about 1 in 500 African Americans have the disease. And as you stated, it affects the red blood cell, which is what carries oxygen around to the body, those cells typically are round and soft and squishy and live 120 days and life is good. In sickle cell, they take a shape like a sickle that you cut wheat with or a banana, and those red blood cells plug up the blood vessels. And that happens anywhere in the body.

As you stated, when that happens in the bones, that's pain. It also happens in the brain, that's a stroke. Can happen in the eye, you lose vision. It can happen in the lungs and cause what's called acute chest syndrome, which can be life threatening. So it's quite dangerous and leads to multi-organ complications and early death for many patients.

**TIM NELSON:** And Rae, you know, you talked a little bit about stigma for, you know, an inherited condition that it seems hard to imagine that. But there seemed to be some association, some cultural associations, with this disease. What's going on there?

**RAE BLAYLARK:** Well, I think, first we have to acknowledge that sickle cell disease is actually the most common inherited blood disorder in the entire world. And, so, that means on basically every continent, you will find sickle cell trait or sickle cell disease. So, the burden of sickle cell disease rests in Africa because of the lower resources.

Sickle cell disease is related to malaria, in the fact, that sickle cell trait is a natural mutation of the red blood cells that really lent itself to survival. So, now, that sickle cell trait and sickle cell disease are ever present, it's really important that there is a universal newborn screening program. It's important that there is education, both in the health care setting with physicians, but also in the community as far as prevalence and transmission and what's in your genes is important for all of us, but particularly, if you carry sickle cell trait.

Because, as Dr. Nelson said, it's a genetic disease, so it's inherited. So, if both parents have this mutation of their red blood cells, whether it be sickle cell trait or sickle cell disease, they stand a chance of having a child with sickle cell disease.

**TIM NELSON:** Now you said it's the most common blood disorder in the world. Why isn't the medical community better at responding to it?

**STEPHEN NELSON:** I'll take that one. I think, some of it may be the history of-- legacy of racism in this country, and discrepancies in funding for diseases that affect the Black community versus other communities.

**TIM NELSON:** And, I'd like to hear, I guess, from both of you, about how you've been fighting that institutional racism and the lack of awareness about the disease.

**RAE BLAYLARK:** Yeah, I'll start with, I think that one of the things that's important is giving individuals who have typically been voiceless, a voice in this space. So, if you're a person living with sickle cell disease, if you're a person who is a caregiver of a child or a loved one with sickle cell disease, it's important that you feel like that there is space for you to share your experiences and talk about what it's like living with the disease.

Having Sickle Cell Foundation of Minnesota, and the fact that, we are a patient advocacy organization, provides this safe space to give individuals a voice, who may have not felt like they had a voice previously. But, we also find ourselves in many spaces of affluence, where we can through our advocacy efforts impact decisions that are made that directly affect the outcomes of individuals living with sickle cell disease.

**TIM NELSON:** Dr. Nelson?

**STEPHEN NELSON:** Yes, thank you for that, Rae. And, I will add to it as a white male physician caring for patients and families affected by sickle cell disease for over 30 years. For the greater part of my career, I was going along thinking I was doing a great job with no awareness how the racial narratives that wash over all of us have been affecting my stereotyping and my implicit bias, which was affecting my cognition and decision making at the bedside. And it really was impacting the care for my patients of color in a negative way.

So, what I have been doing, and I'm partnering with Heather Hackman, who is an educator who does social justice work for educators in developing training for health care providers around this issue of how racial narratives and racism affect our stereotyping and implicit bias, so that we can improve our patient outcomes.

**TIM NELSON:** Now, Dr. Nelson, you talked about being at the bedside. How are people with sickle cell disease treated? Is there an effective treatment for them?

**STEPHEN NELSON:** I guess, there's two ways to address it. There are some effective treatments, sadly, because of some of the discrepancies in funding at the federal and philanthropic levels, with there are only four FDA approved drugs to treat sickle cell disease, so there are some options.

But, even with that, and as you had mentioned, pain is the most common reason why a person with sickle cell disease will seek medical attention, and they are often not treated very well. Their pain is not treated aggressively, and some of that is because there is no physical exam finding or laboratory test to prove that the patient is having pain. We need to believe the patient.

And, now that we are struggling in an opioid epidemic, and the racial narratives that affect our stereotyping, you can imagine that patients with sickle cell disease can have difficulty getting their pain managed effectively.

**TIM NELSON:** I can imagine. But, is there research into longer term approach to this? Are they looking at finding a cure or a cause for this--

**STEPHEN NELSON:** Yes, the answer is yes.

**TIM NELSON:** --is there a way to stop it from passing along genetically?

**STEPHEN NELSON:** Yeah, so there's two-- I'll address your second question first. So with awareness and understanding your status, if you're a carrier for sickle cell disease, you can make some decisions with your eyes open. We would never counsel a couple to not have children. To me that feels reminiscent of eugenics from decades ago. It is important for a couple to understand what their risk is for having a child with sickle cell disease, and there are ongoing trials looking at gene therapy as a curative intervention for sickle cell disease. It is the first molecular disease that was described, and we are excited about the possibility of this cure.

There is a cure available, and that would be stem cell transplant or bone marrow transplant, that's high risk and quite toxic, not everyone has a match. So, the hope and beauty of gene therapy would be that it could be a curative option for every patient.

**RAE BLAYLARK:** I think, it's also important to note that as we are on our way to a cure, that we also have to think about what it means to improve treatment availability. One of those treatments is blood transfusions. Blood is always needed in the sickle cell community, but the matches need to be so close, that it's often that they are looking for donors that are diverse. So, increasing education and awareness around the value and the importance of Black blood donors donating blood and participating in the blood donation process to ensure that we have the best matches available.

**TIM NELSON:** That was Rae Blaylark and Dr. Stephen Nelson. Rae Blaylark is the President and CEO of the Sickle Cell Foundation of Minnesota, and Dr. Stephen Nelson is a pediatric hematologist oncologist and is the Chief Medical Officer at the foundation.