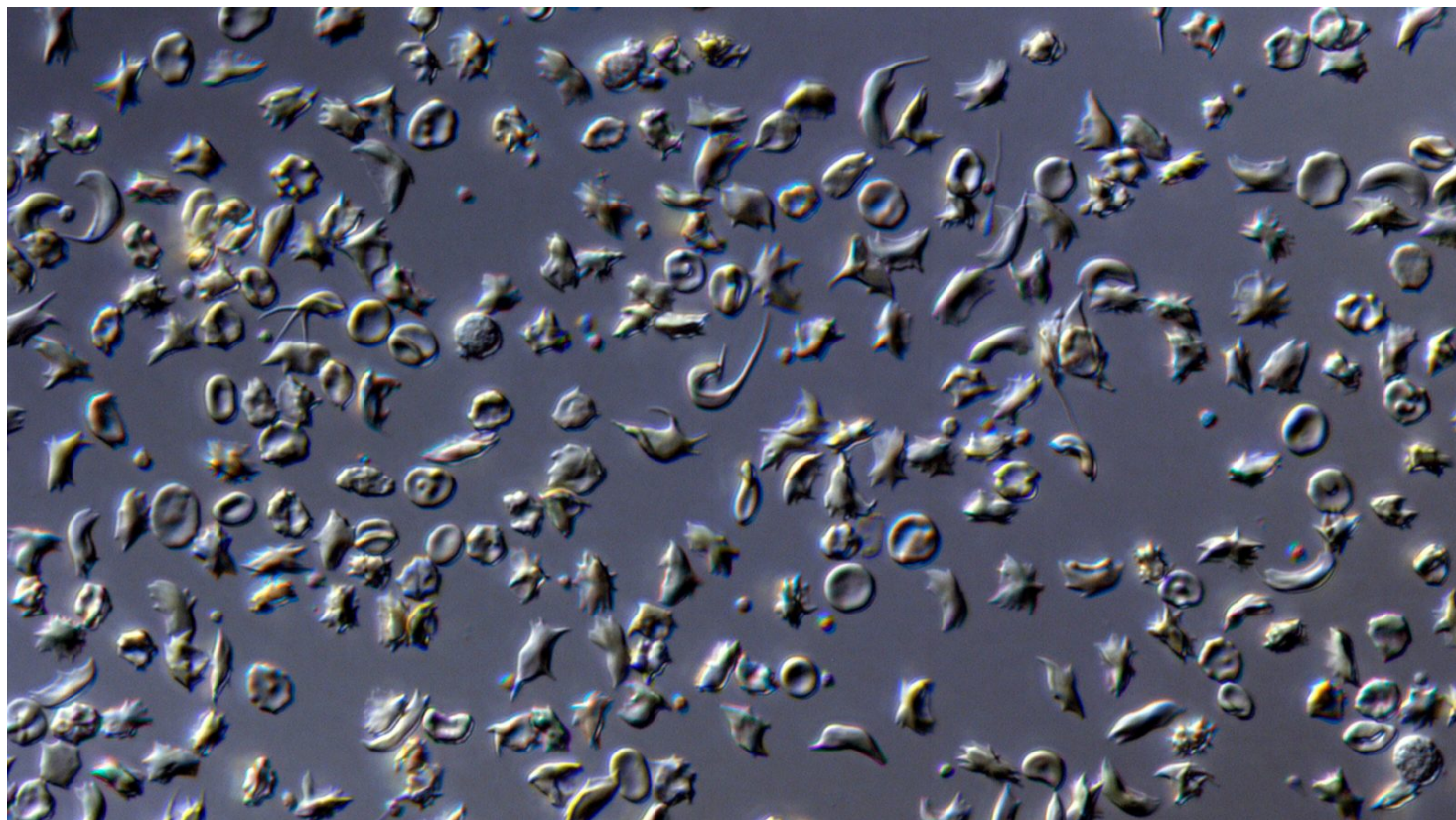


STAT

Sickle cell disease and a pivotal moment to end health inequality

By Ted W. Love July 15, 2021



Red blood cells from a person with sickle cell disease. *Jonathan Armstrong/Wellcome*

Covid-19 laid bare the long-standing vulnerability of minority and low-income communities in U.S. society and its health system even as we celebrated the power of scientific innovation to rein in the pandemic. It's time to turn that innovation, driven by an even greater mind shift, to end inequality in treatment.

In perhaps no condition is such inequality more evident than sickle cell disease — a genetic disease for which the molecular basis has been known [since 1956](#) but for which innovative treatments have been elusive.

Beginning at birth, this disease deforms and destroys red blood cells. Instead of floating easily through the bloodstream as flattened disks, they contort into a rigid crescent or sickle shape. These sickle cells die off sooner than healthy red blood

cells, causing anemia. They can also clump together and block blood vessels, causing pain (known as sickle cell crises or vaso-occlusive crises), organ failure, and early death. People with sickle cell disease die on average [30 years sooner](#) than people without it.

The vast majority of those affected by sickle cell disease in the U.S. are Black and Hispanic Americans, with about 90% of them being Black or of African descent.

Racial bias in the U.S. and its health care system has [left behind people with sickle cell disease](#). They have been mistreated, not believed, not cared for, and often discriminated against in emergency departments and hospitals, including being [stigmatized or doubted](#).

While sickle cell disease has been characterized as a disease of pain, it is in fact a disease of multi-organ failure and premature death. Approximately one-third of people living with sickle cell disease experience two or more extremely painful [vaso-occlusive crises](#) a year. These happen due to the inflammation caused by sickled cells sticking together and blocking blood vessels. Yet the association between sickle cell disease and pain has contributed to the stigma that patients experience. If they aren't in pain, they're given a false sense that they are doing well. Meanwhile, their organ damage advances.

And when they do experience pain, they often wind up in the emergency department or the hospital, and many [share the experience](#) of 33-year-old Cassandra Trimnell. “For many of us, going to the emergency room is a last resort due to the stigma and misunderstanding of the disease. Being dismissed as a drug seeker is unfortunately a rite of passage in our community,” she told me recently.

Decades ago, as a young Black medical school student, I was saddened by how I saw people with sickle cell disease treated, and was embarrassed for the profession I planned to enter. I remember then — more than 30 years ago — wanting to change that.

The situation for Black people with sickle cell disease more broadly reflects the experiences of Black people with U.S. health care: They are often treated

differently than white people and experience worse outcomes. A [2015 review](#) detailed implicit bias against people of color among many white health care professionals. Such bias included viewing Black patients as less intelligent, less able to follow and stick with prescribed treatments, and more likely to engage in risky health behaviors. A [2020 study](#) found that when Black newborns in Florida were cared for by Black physicians, their mortality rate was cut in half, underscoring the lifesaving potential of improving health care for people of color.

The U.S. is now at a pivotal moment for transforming the care of people with sickle cell disease. Why now? First, as a society, we've made strides forward this past year in our social consciousness and ability to talk about racism. And second, scientific innovation may be more appreciated than ever and there have been advances in treating sickle cell disease.

After decades of a dearth in investment and innovation in sickle cell disease, there are now promising treatment options for it. In late 2019, the Food and Drug Administration approved two innovative therapies for sickle cell disease. Oxbryta (voxelotor) tablets, a first-in-class medicine for those with sickle cell disease age 12 and older that directly targets the root cause of the disease, was developed by Global Blood Therapeutics, the company I lead. Adakveo (crizanlizumab-tmca), developed by Novartis, was approved for those with sickle cell disease age 16 and older to help reduce how often painful vaso-occlusive crises occur.

Thanks to a resilient sickle cell community and its researchers, those approvals represent important progress. I'm also encouraged by increased investment and activity in gene therapy, [gene-editing modalities like CRISPR](#), and other innovative approaches to sickle cell disease. More research and more options are positive steps for patients.

But even with accessible treatments, there remain significant gaps in the care of those living with sickle cell disease. Health equality will be achieved only when they are treated with the same level of urgency as white people with rare diseases.

The tools exist to create meaningful change in racial disparities in health care, something that can be done right now for this vulnerable, historically underserved

group of patients. The National Academies of Sciences, Engineering, and Medicine published a [strategic plan](#) in 2020 to help ensure that people with sickle cell disease receive the same high-quality health care to which every American is entitled.

I was raised to believe that the human spirit is grounded by an intrinsic desire to help others. It is time to use this moment to show each other and our children that we acknowledge, but do not accept, the inequality of those in need and then pivot and embrace the knowledge and innovation available to us to make a profound difference.

Ted W. Love, a cardiologist by training, is the president and CEO of Global Blood Therapeutics.

About the Author

Ted W. Love

[@GBT_news](#)

[linkedin.com/in/ted-love-14341711/](https://www.linkedin.com/in/ted-love-14341711/)

 Add a Comment

Create a display name to comment

This name will appear with your comment