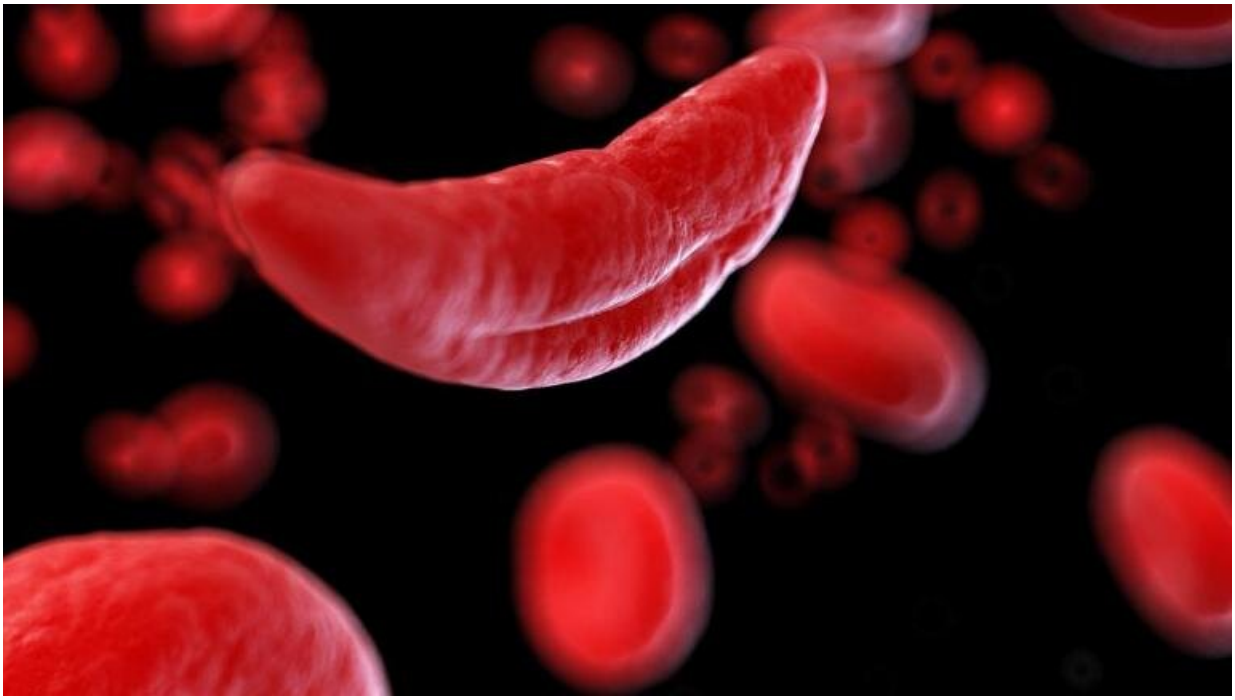
For people with sickle cell disease, ERs can mean life-threatening waits

October 23 2023, by Sara Hutchinson, KFF Health News



Credit: National Institutes of Health

Heather Avant always dresses up when she goes to the emergency room.

"I've been conditioned to act and behave in a very specific way," said Avant. "I try to do my hair. I make sure I shower, have nice clothes. Sometimes I put on my University of Michigan shirt."

It's a strategy to combat discrimination the 42-year-old photographer in Mesquite, Texas, has developed over a lifetime of managing her sickle cell disease, a rare blood disorder that affects an estimated 100,000 Americans. The hereditary condition can affect a person of any race or ethnicity, but Black patients, like Avant, make up the majority of those afflicted in the U.S.

For people living with the disease, a sickle cell crisis can happen at any time. When it does, their rigid, sickle-shaped red blood cells become stuck in their blood vessels, blocking flow and causing extreme pain or breathing difficulties. A crisis can escalate into life-threatening complications such as strokes, seizures, and sepsis.

When a pain crisis can't be managed at home, patients head to the ER to get the high dosage of opioids they need, in addition to IVs to help with dehydration or even blood transfusions. Yet staffers in emergency departments—already overextended and grappling with nursing shortages—don't always have experience in treating the rare disease.

Doctors, amid a still-raging opioid crisis, remain resistant to prescribing the painkillers necessary to treat sickle cell crises. So, patients say, they face long delays before receiving essential care, plus discrimination and suspicion that they are seeking drugs to get high.

"I have to look like I'm not coming in here off the street looking for medication," said Avant. "I have to put on an entire show to get you to believe that I need care."

Years of research have documented the delays. A study published in 2013 found that patients seeking care from 2003 through 2008 at an ER for their sickle cell crises waited 50% longer than patients who arrived with broken legs or arms. A study published in 2021 found that 50% of sickle cell patients reported having to wait at least two hours before their

pain was treated, despite medical guidelines recommending such patients in crisis receive their first dose of pain medication no more than 60 minutes after arriving at the ER.

Medical associations such as the American Society of Hematology, the National Heart, Lung, and Blood Institute, and the Emergency Nurses Association have established guidelines for emergency department-based care of sickle cell pain. And, in 2021, the Emergency Department Sickle Cell Care Coalition, a national collaboration of hematologists, pharmacists, and nurses, helped launch a point-of-care tool to help medical professionals manage the disease in the ER.

But patients and sickle cell experts said those best practices haven't been widely adopted. A 2020 survey of nearly 250 emergency medicine providers found that 75% of them were unaware of the NHLBI's recommendations, first published in 2014, yet 98% felt confident in their ability to treat patients with sickle cell disease.

Still, ER horror stories abound among adults with sickle cell disease. For Lesly Chavez, 29, a Houston hairstylist, her worst experience occurred a few years ago. She said she spent four hours in a waiting room before getting seen.

"And when they finally got to me, they told me they could help with 'my addiction," but they decided that there was nothing that they could do for me," Chavez said. "They just flat-out said no and sent me home while I was in crisis."

Chavez said she has since avoided that hospital even though it's 10 minutes from her home. Now she drives to an ER 30 minutes away.

Chavez, who is Hispanic, said she confronts "doubt everywhere I go" because sickle cell disease primarily affects Black Americans. (Those

who are Hispanic can be of any race.)

Paula Tanabe, a professor of nursing at Duke University who has spent decades researching ways to improve care for sickle cell patients, said a confluence of factors adds to the racial bias patients may face.

"Emergency rooms are incredibly overcrowded, at rates that we have never seen before, and that's for everyone," said Tanabe.

Legislators are trying to help. A federal bill introduced in June would allocate annual funding for five years to a program that trains doctors on best practices for caring for sickle cell patients. Another, introduced this spring, would provide funding for community organizations working to spread awareness about the condition and give student loan relief to medical providers who commit to working on the disease. Some state legislatures have established sickle cell task forces to improve physician education and care coordination.

Advocates for sickle cell patients said investment in data collection to track the disease is also important. Although the Centers for Disease Control and Prevention estimates that some 100,000 Americans have it, the true number is unknown. That's because no national system exists to collect data on sickle cell, unlike other conditions such as diabetes, cancer, and Alzheimer's.

"I'm 32 and we've been saying it's 100,000 my entire life," said Quannecia McCruse, who co-founded the Sickle Cell Association of Houston. "I know there's more. I know people are going uncounted."

Eleven state-led data collection programs currently exist and, in February, the CDC opened a new grant application for additional states. Improved data would allow funding to be allocated toward the areas with the greatest need, sickle cell patient advocates said.

Texas had an opportunity to join those efforts. This spring, the state legislature passed a bill with broad bipartisan support to create a sickle cell patient registry, but Republican Gov. Greg Abbott vetoed it, saying it would compromise patient privacy.

"That was a bad excuse," said McCruse. "We have a cancer registry already, and everyone's information is safe. That registry would have gone a long way to help."

While progress grinds slowly, patients like McCruse say they're forced to balance advocating for themselves during bouts of excruciating pain against the need not to irritate or alienate hospital staffers.

"It feels like someone is taking a Taser and shocking the crap out of me. Or when it's really bad, and it feels like shards of glass are just moving through my veins," said the mother of two. "It's very, very painful. And you're telling somebody whose body is torturing them that it's not that bad?"

Alexis Thompson, a hematologist who treats sickle cell patients at the Children's Hospital of Philadelphia, said she works with her pediatric patients to develop self-advocacy skills. But sometimes that backfires.

"The great irony is patients who are well informed and capable of self-advocating are being accused of being manipulative, because they are capable of articulating very clearly what's effective for them down to the name of the medication or the absolute dose," Thompson said.

Sickle cell experts recommend that doctors adhere to a patient's individual pain plan, if available. Thompson said those plans, which document patients' diagnoses alongside a recommended medication and dosage, can be uploaded to online portals that patients can pull up on their cellphones when visiting an ER to verify what they need.

Patients such as Avant hope such steps can help decrease their ER waits while easing their anxiety about seeking emergency care.