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STAT

# ‘Every time it’s a battle’: In excruciating pain, sickle cell patients are shunted aside

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Sept. 18, 2017

Amy Mason, who has sickle cell disease, at her home in Mobile, Ala. “Every time, it’s a battle,” she says of seeking care in the emergency room.
*Sharon Steinmann for STAT*

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Amy Mason had toughed it out for hours one day this past July, trying warm soaks and heating pads and deep breathing to soothe pain that felt like her bones were being sawed with a rusty blade.

She knew this was a life-threatening emergency of sickle cell disease, in which her misshapen red blood cells were getting stuck in her blood vessels like tree limbs in a storm sewer. But she delayed going to the emergency room; previous visits hadn’t gone well.

Just before midnight, Mason, 34, finally had her boyfriend drive her to a Mobile, Ala., hospital. She told the triage nurse that she was having one of the worst sickle cell crises of her life and that she was off the far end of the 1-to-10 pain scale. She was told to wait.

As the hours passed, Mason begged her boyfriend to take her to another hospital, then passed out from the pain. She awoke, she told STAT, to her boyfriend’s shaking her and pleading with her to stay with him. *I can’t do this anymore*, Mason whispered.

Daylight broke. Around mid-morning, an ER nurse told her, *you know, we’re not a pain clinic*. She gave Mason a small dose of the narcotic Dilaudid.

"Every time, it's a battle," Mason said about the ER, where the staff suspected her of faking her condition in order to score opiates and viewed her as a non-emergency because she "only" had pain. "Nobody cares about people with sickle cell."

The U.S. health care system is killing adults with sickle cell disease. Racism is a factor — most of the 100,000 U.S. patients with the genetic disorder are African-American — and so is inadequate training of doctors and nurses. And the care is getting worse, sickle cell patients and their doctors said, because the opioid addiction crisis has made ER doctors extremely reluctant to prescribe pain pills.

STAT interviewed 12 sickle cell patients who described the care they received and didn't receive. They were old and young, men and women, scattered from coast to coast, some with jobs or attending school and some too sick to do either. Two who wanted to tell their stories were unexpectedly hospitalized and too weak to talk to a reporter.

In hospitals, sickle cell patients are typically treated by generalists who know little about the disease and patients' desperate need for pain relief. At one hospital, researchers found that sickle cell patients waited 60 percent longer to get pain medication than other patients who reported less severe pain and were triaged into a less serious category.

Outside the hospital, fewer than one-quarter of adult patients who need it get the drug approved for preventing sickle cell crisis, according to the American Society of Hematology.

Only 20 percent of family physicians say they feel comfortable treating sickle cell disease, a 2015 survey of more than 3,000 such physicians found, leaving many patients without routine, preventive care. They are therefore more likely to seek care in ERs during an acute sickle cell episode, called a vaso-occlusive crisis, in which an inadequate blood supply triggers excruciating pain and damages vital organs.

The result of all these failures is "a breakdown in the system of care for these patients," said Marsha Treadwell, of Children's Hospital Oakland, who works on programs for adults with sickle cell. "People are needlessly dying."

And they are dying in droves, of vaso-occlusive crises and organ failure and chronic kidney disease and more. The ultimate cause is eponymous sickle-shaped blood cells. They don't deliver oxygen to organs and tissues as healthy blood cells do, causing slow, inexorable damage over decades.

But an immediate cause of early death is a catastrophic failure to give sickle cell patients competent care. The mortality rate for adults with the disease has risen 1 percent every year since 1979, the hematology society reported in 2016. Half of adult sickle cell patients are dead by their early 40s. (Children fare better.)

Every doctor who cares for adult sickle cell patients "knows of one or more who were not treated properly and died," said Dr. Keith Quirolo, a retired pediatric hematologist in California who saw adult patients because no other physicians would accept them.

Each and every one of the patients STAT reached, through doctors and social media, sounded exhausted, even defeated. Some spoke of hospital and other committees they had served on, only to see the treatment guidelines and educational materials they produced ignored. Most spoke about fellow patients who'd recently died — a man in his 30s, a woman in her 40s — and who might have been saved with competent care.

By the time Mason was given pain medication during her July episode, she said she was struggling to breathe. She was too weak to even boost herself onto a bedpan; her boyfriend lifted her. He had to buttonhole a doctor and insist he treat Mason.

The Mobile Infirmary hospital, where Mason was treated, said it couldn't comment on the care of a specific patient. Registered nurse Kelley Hicks, director of emergency services, said, "We do our best" to give a bed to "each patient that arrives to the ED so that their care may begin as soon as possible. … Our goal is to always provide high quality, evidence-based care to all of our patients."

But Mason felt they fell short. "You go into the emergency room and you don't know if you're going to leave alive," said Mason, who lost two friends to the disease this summer. "We are dying way too young."


The emergency room at the Mobile Infirmary hospital in Mobile, Ala., where Mason was treated in July. *Sharon Steinmann for STAT*

## 'I always dress professionally'

Physicians are taught that signs of drug seeking include asking for opioid medications by name, making multiple visits for the same complaint, having symptoms "out of proportion" to what an examination shows, and seeking treatment at a hospital. That means four strikes against patients seeking care for acute sickle cell crises in an ER.

The opioid crisis has made physicians even more vigilant and cautious, as everyone from the Centers for Disease Control and Prevention to state regulators to some hospitals have tightened guidelines for dispensing the addictive pain medications.

Racial bias makes matters worse. Numerous studies have found that black Americans receive less pain treatment than whites, often because physicians and nurses hold false beliefs such as "black skin is thicker."

"If you are African-American and are in the ED for pain, you are almost automatically tagged as drug seeking," Quirolo said. In one study, 63 percent of nurses surveyed said many patients with sickle cell are addicted to opioids. In fact, their rates of addiction are no higher than the general population's, said Dr. Alexis Thompson, president of the hematology society. They may even be lower: Although blacks make up 13 percent of Americans, they accounted for only 8 percent of opioid overdose deaths in 2015, the most recent year studied.

Knowing the attitudes they'll encounter, patients therefore prepare for the ER as for a job interview. "I always dress professionally," said Wanda Williams, 67, a retired school administrator in the Bay Area: nice shoes, "interesting earrings," every hair in place, and Vogue-worthy makeup. "It's crazy that you, as an African-American, have to do this so you aren't treated like a drug addict."

Such efforts to increase the chance of receiving appropriate treatment often fall short. Several ER physicians told STAT that everything would go fine if sickle cell patients brought their "care plan" — a document from their doctor showing their diagnosis, and what drugs and intravenous hydration and possibly transfusions they should receive in a sickle cell crisis. "But what we find is that emergency physicians and hospitalists [who treat inpatients] don't like to follow these plans," said social worker Kim Major, who oversees the sickle cell clinic at Oakland Children's.

The plans' descriptions of pain medication instead raise suspicions. "We want to have informed, empowered patients, so we've taught them to be aware of what their medical needs are, what they take, and at what dose," said Thompson. But knowing so much in such detail, she said, "is perceived as drug seeking."

James Griffin, 36, knows that if he suffers a sickle cell crisis on a Saturday night, when ER doctors are extra suspicions of black men arriving in need of pain medications, he needs to hold on until morning. He also knows to be cooperative even at a sacrifice to his health.

"If the doctor suggests one pain medication, even when it's one that I had a respiratory reaction to, I don't suggest something else," said Griffin, who lives in Phoenix and is training to become a medical assistant. "If I do, I might not get anything" because doctors are suspicious of patients who know "too much" about what pain medication they need, he said.

Emergency physician Dr. Jon Hirshon of the University of Maryland explains the wariness. "When a patient tells me that this dose of this pain medicine works for them, I need to have some type of objective information to support that," he said. "If someone comes in off the street I don't know what that's based on." There is no objective measure of pain, and because sickle cell patients build up tolerance to medication, their doses can shock physicians.

Nevertheless, Hirshon said, "The vast majority of the individuals with sickle cell disease who go to the emergency department get excellent care."

He and other emergency physicians feel they are in an impossible position, made worse by the opioid crisis and the resulting crackdown on irresponsible prescribing of the addictive drugs. "A lot of sickle patients are chronic pain patients, and state guidelines say you don't treat chronic pain in the ED," said Dr. Howard Mell, an emergency medicine physician in Ohio and spokesman for the American College of Emergency Physicians. "These patients are like most humans: Some of them are addicts. I have to verify the need for these powerful medications."

If he makes the wrong call, Mell said, "I feel genuinely awful: 99 out of 100 [sickle cell] patients will have a legitimate need [for the pain medication they request], but the 1 in 100 could cost me my license." While state regulators would be unlikely to discipline a doctor for one such mistake, many physicians voice this fear.

Mell tries to determine whether the pain is genuine, he said: "If I don't know this patient, and she tells me her knees and ankles are killing her and this is her typical sickle cell crisis, if there's no increase in blood pressure or pulse, I'll question it. Emergency physicians are pretty good at sussing this out."

## Fatal consequences

Providing competent treatment in the ER is about more than alleviating suffering. If patients in sickle cell crisis don't get treated with appropriate quantities of fluids, medication, oxygen, and more, the

consequences could be fatal.

Yet, ignorance of sickle cell among emergency nurses and physicians is well-documented. In one study led by emergency nurse Paula Tanabe of Duke University, the average score on a test of basic emergency care for sickle cell patients was 65.

Adrienne Shapiro, an advocate for patients with sickle cell disease in California, said she gets frequent calls like this from patients in the ER or their families: Come as quickly as you can. Last February, it was Natasha, sobbing.

Guidelines from the National Institutes of Health, in effect since 2014, call for patients to be given pain medication within an hour of registering in the emergency department, with additional doses until the pain is controlled. Natasha (whose family asked that she not be further identified) had not received any medication during the five hours and counting since she had arrived at an ER in southern California, nor had she been given fluids or oxygen.

Asked by Shapiro why Natasha, in her 40s, had not been treated, the ER physician said it was because Natasha had been there the day before, also complaining of pain.

“You do understand that she has sickle cell?” Shapiro asked. The doctor was adamant that she would treat Natasha’s pain only if she also agreed to a blood transfusion as an indication that she really had sickle cell disease.

Because Natasha had had so many transfusions, Shapiro explained to the doctor, any more could cause a fatal immune response. Natasha was eventually treated with fluids and pain medication and sent home.

Two days later she was admitted to another hospital, Shapiro said. Within 18 hours, she was dead of sepsis, a systemic inflammatory response to infection that is common in sickle cell. It’s impossible to know if she would have survived with faster ER care, but pain causes the blood vessels to constrict, exacerbating the oxygen deprivation to vital organs caused by the disease.

### ‘There’s no money in it’

There is no surefire way to avoid sickle cell crisis and therefore the ER, but receiving routine care helps. That is easier said than done.

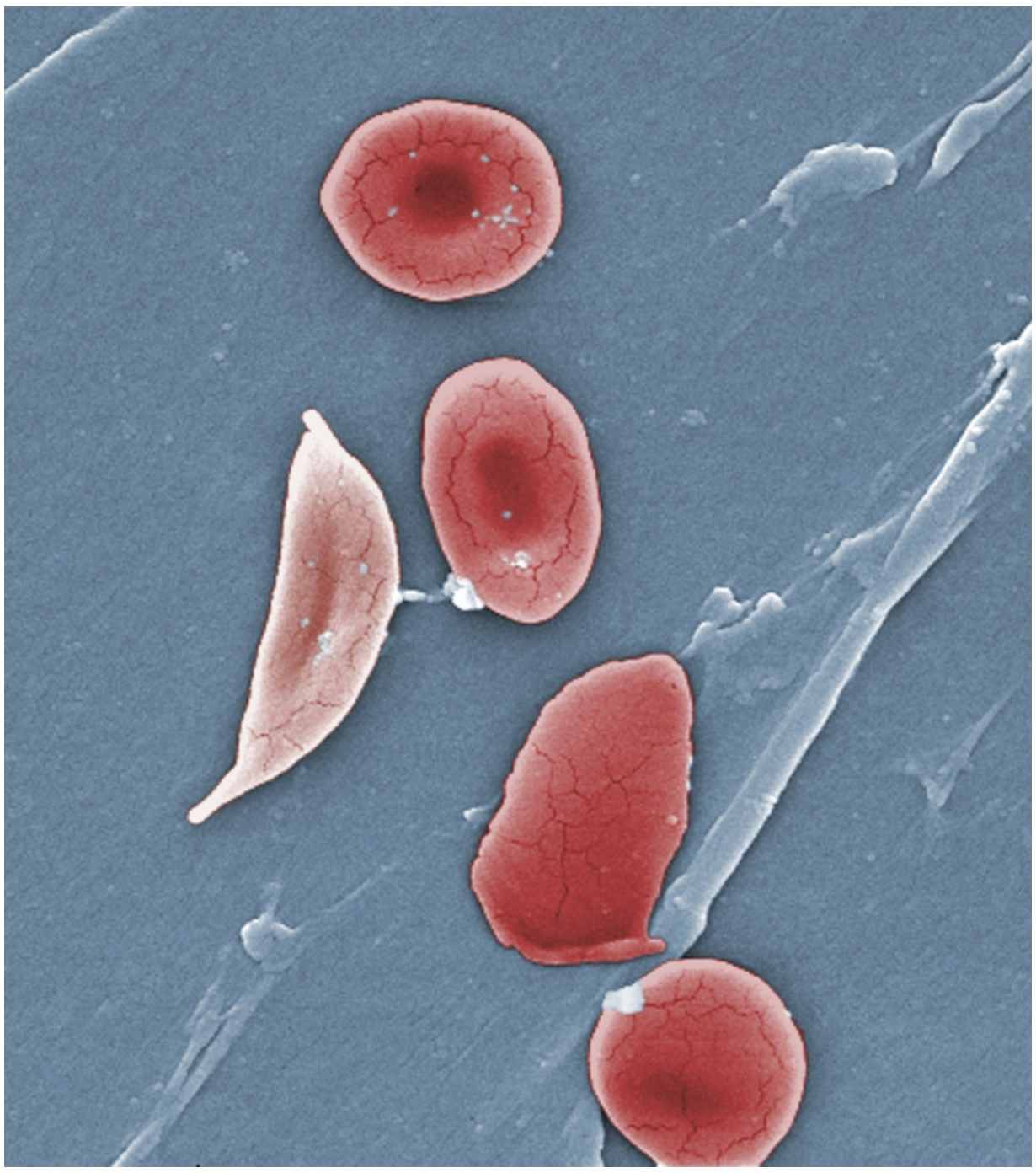
“We don’t know what percentage of sickle cell patients have an adequate primary care provider,” said Thompson, the hematology society president. “But it’s safe to say a substantial number don’t.”

In part, that’s because sickle cell disease is complicated. Caring for patients requires knowledge of, and access to specialists in, cardiology, pulmonology, nephrology, and more. But another reason is that “most primary care doctors don’t know how to take care of these patients,” said Duke’s Tanabe. Medical schools devote an hour or two to sickle cell disease; unless they train in a city, interns and residents might never see a case.

“That immediately puts into the mind of our future doctors and nurses, OK, this isn’t important,” said Marissa Cors, 40, a patient in southern California. “They don’t take it seriously. I once heard a doctor say, ‘Sickle cell is said to cause great pain’ — as if that were a rumor we started and not a medical fact.”

Cassandra Trimnell, 30, president of the education and patient-support site Sickle Cell 101, has had doctors ask her, “How long have you had sickle cell?” As an inherited disorder, it is present from birth.

Even the specialists who are supposed to treat sickle cell, hematologists, often don’t want these patients. “If you’re a hematologist, there’s no money



A sickle cell (left) and normal red blood cells of a patient with sickle cell disease. *Janice Haney Carr/CDC/Sickle Cell Foundation of Georgia via AP*

in it," Quirolo said. "The patients are complicated and hard to take care of, and it's very time-consuming." Only about five physicians in California, which has an estimated 7,000 sickle cell patients, specialize in treating adult sickle cell patients, he said, "and this is the way it is across the country."

For hematologists, treating blood cancers such as leukemia is much more lucrative. "Let's be honest, that's where the money is," said Dr. Jane Hankins, a sickle cell expert and hematologist at St. Jude Children's Research Hospital in Memphis. "And hematology training isn't focused on sickle cell disease."

Wanda Williams tried to get treated at a center that specializes in sickle cell, but her insurer told her she had to instead choose an in-network hematologist.

"I called and described my situation, and they said we don't know what to do with you," she recalled. "I've had doctors say, 'I don't take care of sickle cell patients. They're too much work, and they die.' I have to sell myself in order to get someone to take me as a patient." She now drives 30 miles each way to a hematologist.

One benefit of routine care from a physician who knows her way around sickle cell disease is receiving hydroxyurea, arguably the most significant breakthrough in sickle cell treatment ever. The oral drug increases healthy forms of oxygen-carrying hemoglobin, resulting in fewer pain crises, fewer transfusions, fewer emergencies, and less organ damage.

But many physicians are leery of hydroxyurea. It was originally, and still is, a cancer drug. It can reduce the number of white blood cells, so patients require blood tests every month or two. To work well, it must be given at the maximum tolerated dose, which patients build up to over about six months.

"It's not a rocket science medication to manage," said Treadwell, of Oakland Children's. "But primary care physicians just don't want to prescribe it."

As a result, sickle cell patients who might have avoided vaso-occlusive crises and the ER wind up there and, often, as hospital inpatients.

There, they are typically treated by hospitalists, a specialty created about 20 years ago so physicians wouldn't have to leave their offices to care for their hospitalized patients.

"Hospitalists know a little bit about everything, but with sickle cell, we're talking about a multi-organ disease," said Yvonne Carroll, director of patient services in the hematology department at St. Jude.

After her last vaso-occlusive crisis, Trimnell was admitted to a Florida hospital. Her mother told the hospitalist assigned to her that Trimnell needed to be transfused, but he refused, saying he didn't understand how a transfusion would help. It's a standard intervention.

"I was left for days with my hemoglobin dropping," Trimnell said. Her doctor in California phoned the hospitalist at 3 a.m. explaining the need for a transfusion, which she finally received. "If he hadn't insisted I would have died," she said.

A number of efforts are underway to fix a system that is letting patient after sickle cell patient die. Goals include improving ER care, including by educating staff about the disease. But patients and doctors alike told STAT that the failings in clinical care won't be solved without addressing racial prejudice.

"I just think," said Duke's Tanabe, "you can figure out a way to take care of sickle cell patients who come to the emergency department, once you get past the attitude of 'they're black and they're addicts.'"

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