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Sickle cell disease is complex on its own, but black men with the illness battle its stigmas and stereotypes too

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Doctors didn't expect Marqus Valentine to live past age 5. The prognosis was so certain that the Valentine family was granted a free trip to Universal Studios to the set of "The Ghostbusters" from a foundation as a dying wish.

Although Valentine beat those odds, his troubles didn't stop. Diagnosed with sickle cell disease as a baby, he repeated kindergarten because of missing a lot of school due to lengthy hospitalizations and even had a stroke in fourth grade during class, he said.

Growing up during the height of the AIDS epidemic, his classmates didn't want to play with him because they thought Valentine was contagious. Invitations to birthday parties stopped, and as he got older, he spent the rest of his childhood years receiving monthly blood transfusions, as well as other treatments.

"My peers, no one understood sickle cell," said Valentine.

The disease is complex physiologically, manifesting itself differently in every carrier, but also sociologically, bringing in factors like racial bias, since sickle cell is believed to be most common among minority groups, particularly African Americans. Historically, there has been limited funding and research dedicated to sickle cell disease, evidenced by only one FDA-approved drug available to the community until last month.

The landscape for adults who have sickle cell is much harder than that for children with the illness, and black men specifically may have a harder time dealing with the condition because of existing stereotypes preceding them. A lack of resources for a disease that primarily affects a marginalized community, coupled with negative perceptions of African American males, can exacerbate the navigation of an already complicated, painful disease.

Sickle cell disease, given it's name for the abnormal C-shape the red blood cells take—similar to a sickle farm tool or a crescent moon—occurs when there's a genetic mutation in a person's hemoglobin, the protein source in red blood cells responsible for carrying oxygen throughout the blood, according to the American Society of Hematology.

There are different types of sickle cell disease, which is inherited when a child receives two sickle cell genes—one from each parent, according to the Centers for Disease Control and Prevention.

In the United States, the exact number of people living with sickle cell is unknown, but the CDC estimates that 100,000 people in the country are affected by the disease. One out of 365 African Americans have sickle cell disease, and 1 in 13 are born with the trait. For Hispanic Americans, the prevalence is about 1 out of every 16,300.

TaLana Hughes, executive director of Sickle Cell Disease Association of Illinois, said there are only approximate numbers because there is not an official registry for sickle cell disease.

"(For a while) they've used that there are 100,000 people in the U.S. that have sickle cell disease," said Hughes. "Well, wouldn't we think that that should be larger now?"

The issue with this, explained Hughes, is that there is no registry where,

upon diagnosis, typically as a baby, you're entered, preventing an accurate accumulation of data and long-term follow-up, she said.

Illinois added sickle cell disease as one of the conditions to be tested for in the newborn screening panel in 1989, according to data from the Illinois Department of Public Health. Since then, the state has maintained legislation around the disorder, Hughes said. There are plans for funds to be allocated to a universal sickle cell registry, a more accurate numerical database of people living with the disorder, she said. "We still kind of have to just wait," she said.

In September, the CDC awarded funding to seven states, the closest to Illinois being Indiana, to work with the CDC on a data collection program to start accumulating comprehensive health information on people with sickle cell disease throughout the next year. A total of nine states are in the program.

Hughes, who's been in her role as director for almost a decade and who also has a 17-year-old daughter with sickle cell disease, said she spends a fair amount of her job fundraising and securing grants since her organization does not receive any state funding.

"We're advocating all the time," said Hughes, who's been at the organization since 2002. She feels that the stigma of sickle cell being "a black disease" is part of the reason it doesn't get as much funding or attention.

"Over the years, instead of (sickle cell) having the description of being a genetic disorder, because that's what it is—meaning, genetically anybody can pass it down—it's been called a 'black disease'," she said. "It's like, 'Oh, that's a black problem so the black people need to figure out how to fix that,' kind of thing."

Dr. Lewis Hsu, a pediatric hematologist at UI Health, the University of Illinois at Chicago's health system, explained that the disease is incredibly complicated for its physical and psychosocial pains, but also for its ties to "all kinds of issues with social justice, race disparities and stigmas."

"The amount of problems that arise because this (disease) affects minorities is huge," he said.

One of the hallmark problems of sickle cell disease is pain: a "pain you can't see from outside," Hsu said.

"The pain can be stupendously severe," he said, "worse than a broken leg, or worse than childbirth; it's a 10 on a scale of 10. It happens unexpectedly and for no clear reason on when it will stop."

Until recently, there hasn't been much hope for cures or treatment, said Hsu, only palliation methods to ease the pain. So when sickle cell patients experience a pain crisis, they often go to the emergency room seeking relief, sometimes resulting in accusations of being drug addicts.

Valentine, now 36, felt hurt when this happened to him, especially since the accusation came from a nurse who had taken care of him several times in the past, he said.

"I asked for my IV medicine even though I had it written for IV, oral and I was on a pain pump," he said. "For a nurse, who has taken care of me in the past, to come in to my room and say, 'I'm going to not, you have oral written,' I was shaken up, sad and I wanted to get angry but I couldn't."

"He still required additional IV pain medication doses despite being started on the oral medications," elaborated Ashley Valentine, 31,

Marqus' younger sister, who said her brother had just finished spending a prolonged time in the intensive care unit for severe sepsis prior to this interaction. "Marqus was in distress and called (me and my mother) because he was told that he would not be receiving IV pain medication from 'this nurse' during 'my shift.'"

This was against Marqus' established pain plan covered by hematology and anesthesia pain service, explained Ashley, who works full time as president of Sick Cells, the advocacy nonprofit organization that she and Marqus co-founded.

"As a man, it was heartbreaking because I was there u2015 I didn't want to be there—I went from being super healthy to now being stuck in a hospital room dying," said Marqus Valentine.

Situations like these are not uncommon and one of the main stigmas black men with sickle cell disease face when seeking emergency medical attention for a pain episode, said Hsu.

"If someone with sickle cell disease goes to an urban hospital, and you're an urban African American male, and people just look at you like, 'Yeah, right, you just want these meds," said Hsu, "it's instant stress (for the patient). Even when seeking help, when people are in helping professions, there's raised eyebrows and skepticism. There is, 'No, we don't believe you. Prove you have sickle cell disease.'"

Men may face issues like these more than women do, said Hsu, due to stereotypes and ways of communication distinct to black males. He trains his teenage patients through role playing on how to deal with these scenarios, but the navigation can be harder for adult males.

"The times I do get sick and have to go to the emergency room, if there is staff who don't know who I am, it is hard," said Valentine.

Elijah Powell, a 24-year-old Hyde Park native who was diagnosed with sickle cell disease at 2 months old, said he thinks the stigma of drug addiction for sickle cell patients applies to black men more.

"Look at all the stigmas a normal black man faces on a regular basis," he said, "then you add sickle cell to it and it's way more extreme."

Powell, who graduated from Morehouse College in Atlanta with a biology degree in 2017, said it's hard having sickle cell disease as a young black man because of how the condition depletes the body.

"The illness itself drains you and naturally makes you weaker," said Powell, who later mentioned he's not as masculine as he'd like to be, a common sentiment among men with the illness, according to Hsu. "Most black men, they're strong and able to do a lot of stuff, but we can't do as much because our body is weak and our organs are breaking down and being attacked by sickle cell disease on a daily basis."

Powell works as a freshman on-track coordinator at Kenwood Academy High School. He says he mentors youth with sickle cell disease so they can have hope.

"I try to show sickle cell patients you can go to college, pursue your dream job and live a life you've dreamed of," said Powell.

"I'm hoping for a cure one day," he added, "but we'll see what happens."

There have been new developments for curative treatment options for adults involving gene therapy treatments, like stem cell transplants.

"Ever since people learned this was a genetic condition, we knew there was a possibility of doing a genetic cure," said Hsu, who says the techniques still have to be proven, but that there is considerable promise.

After battling sickle cell his whole life, Terrance Hill, a 40-year-old West Pullman resident, received a successful stem cell transplant in 2016, he said, leaving him with just the trait.

"I got the half cure," he explained, meaning he's left with the trait as opposed to being completely sickle cell free. For the latter, he needed to get a donor who was a full, identical match, he said. Hill was able to get the transplant because his doctor said he might be able to benefit from the treatment since he wasn't having too many complications or crisis at the time.

Since the transplant, Hill says he's "been OK," and that living with the trait has been "somewhat better" since he's able to do more physically.

"I'm moving around, exercising more, just able to participate in different events," he said. "Beforehand, it was just hard to do things; just trying to come out was just hard. It took a toll on my quality of life."

In November, just 10 days apart, the FDA approved two new drugs: Adakveo, which would reduce the common pain crises, and Oxbryta, which would make red blood cells less likely to form the sickle shape.

Ashley Valentine called these drugs "game changing," because "these are the ones that actually, specifically target sickle cell."

"This is such a big deal because up until this point there has not been investment into sickle cell disease, and a lot of that is because of the population it impacts," she said. "The disease was discovered in 1910, and it's taken this long for the game-changing drugs to come."

Prior to these two approvals, there was only one medication for sickle cell called Hydroxyurea that was approved by the FDA in 1998, and it was specifically for adults, according to data from a 2016 report from

the American Society of Hematology.

The new medications are exciting for the community, Hughes said, but there is still some hesitation.

"Unfortunately, we went through decades of no treatment options," said Hughes. "When you have one thing that's being offered as an FDA approved treatment option—and say, for instance, that doesn't work because we know that all medications don't work for some, or some don't work for everybody—you kind of have a community that hope has been lost because for so long there's been no options."

Additionally, Hughes explained how adult patients, who've had much wear and tear on their bodies from years of harsh pain medications and blood transfusions—that only treat symptoms—may not have the physical wherewithal to stand a new treatment or even a curative option.

"You still have people dying from sickle cell disease," she said. "With blood transfusions, over time, you can build up antibodies. If you're taking these harsh pain medications for 20 years, you know long term there's going to be damage to your organs. There really weren't things (in the past) put in place to help treat your body."

Marqus Valentine, who has been on Hydroxyurea since he transitioned to adult care, will start Oxbyrta this month and is optimistic. He'll keep a journal to chart how his body responds to the medication, he said.

"Going from (what felt like) nothing to two drugs specifically designed for sickle cell is excellent," he said. "It's been kind of like a weight off my shoulders."