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Crisis Interventions

Researchers are searching for new ways to help kids with sickle cell disease.

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Comedian Kier Spates recalls being a sickly child. He was constantly ill, often wracked with pain, and sometimes he screamed all night long. But his parents didn’t know why. The pediatricians thought Spates was reacting to allergies

Finally, when he was 7, a doctor diagnosed him with sickle cell disease (SCD). That’s when Spates’s journey with this red blood cell disorder began.

If someone has sickle-cell anemia, some of their normally O-shaped red blood cells are C-shaped. These sickled cells can cluster and create blockages in small arteries and capillaries, stopping blood flow and causing pain. “For seven years, all my parents knew was I was hurting,” Spates says.

After his diagnosis, Spates continued living life normally. “At 7 years old I really didn’t think I was different from other children,” he says. “I still played and did the things that I loved to do; just sometimes I had to go to the hospital.”

But as the years rolled by, Spates learned just how devastating SCD could be. As a teenager, he was an avid athlete. Spates ran track and played basketball and football. “But someone mentioned to the football team doctor that I had sickle cell, and he threw me off the team,” he says. “My only dream was to play in the NFL, and I had that snatched away from me because of sickle cell.”

Spates sunk into a depression. “I was good at football and I just couldn’t understand why I couldn’t play anymore,” he says. “I didn’t go around the team anymore, I didn’t come out of my room, and I didn’t talk to my parents. I didn’t know what to do.”

Keeping their son healthy also strained and finally derailed Spates’s parents’ marriage, further deepening his depression. He attempted suicide twice. But Spates rallied. “I really think about that moment and those days, and it keeps me going,” he says. “I don’t ever want to be in that mental state again. So I started educating myself about the disease. I told myself that I had to face my fears.”

This change in attitude helped Spates better manage his disease. He vowed to stay positive and to work with his doctors to improve his quality of life. “It’s so important that you understand you can have sickle cell and still live a full, quality life,” he says.

When Spates was diagnosed with SCD in his hometown of Houston, like many states, Texas didn’t have mandatory testing of infants for sickle cell disease. But on May 1, 2006, the federal government required all states to conduct universal screenings of newborns for SCD.

This paved the way for national health organizations to establish clinical guidelines to treat sickle cell disease in children. “By and large, the programs developed to manage children with sickle cell disease in this country are good,” says W. Keith Hoots, MD, director of the Division of Blood Disease and Resources (DBDR) at the National Heart, Lung, and Blood Institute (NHLBI). DBDR’s mission is to support and develop research and training programs on the causes and prevention of blood diseases and disorders, including SCD.

Hoots says the multidisciplinary programs help children living with sickle cell disease and their parents manage the physical, psychological and social stresses caused by SCD. They help kids integrate into schools and other settings that are necessary for their growth. “All those things are part and parcel of what some of the best programs do for children,” he says.

Kier Spates

But Spates grew up during a time when SCD was largely misunderstood. Doctors predicted he wouldn’t live past the age of 11. His response—with the love, commitment and understanding of his parents and a physician who was a friend of the family—was to prove them wrong. “My mama told me, ‘You can do anything you want to,’” he says. “I vowed never again to let sickle cell disease stop me from doing anything.”

At age 19, Spates, who had a gift for making people laugh since he was a child, decided to become a comedian full-time. Today, the 37-year-old entertainer works on the popular syndicated radio program The Steve Harvey Morning Show. Spates is also the celebrity ambassador for the Sickle Cell Disease Association of America (SCDAA) and is a spokesperson for the agency’s “Rise Above” campaign. “Our goal is simply to raise awareness about sickle cell disease as much as we can and where we can,” he says. “As long as sickle cell has been around, understanding of the disease is still in its infancy, and we have to get people talking. We are the greatest country on Earth and it’s time for us to do something about this disease. My biggest goal is to draw as many people into this fight as I can.”

Spates also launched his own foundation, Kier’s Hope, to help families dealing with sickle cell. Spates says the big goal for his foundation is to organize the first-ever “sickle-cell family reunion.” “I think it is very important that we treat this disease as a national issue, and we would like to call attention to it through this event, a family reunion in one city. We're all bound by blood, and that makes us family," he says. "People with SCD don't get to have a lot of fun. We spend so much time in the hospital, in doctors' offices, getting shots, giving blood, getting blood transfusions; wouldn't it be nice to have a day where we can just sit back and relax and enjoy some music, socialize with each other and just have fun?"

On the medical side, Hoots and other researchers are working hard to establish a continuum of health care for kids living with SCD as they transition into adulthood. One huge concern he has is that as children age out of pediatric care, the support systems that help manage their sickle cell disease tend to disappear. In part, this is because of the way reimbursement for medical services is structured, and partly because the health system puts more responsibility on individuals with SCD moving into adulthood to manage their own care, he explains.

"That's appropriate, except when you have a disease that must be managed every day," Hoots says. "That's not only exhaustive and exhausting from a health care perspective, but exhausting in terms of economics. Imagine explaining to everyone why you can't show up to your job today because you're in pain and just incapacitated. Just as these things can happen on the health care side, they can happen in the wider arena [of life]."

Hoots believes one way to help kids with sickle cell prepare to manage the disease when they move into adulthood is to teach them how to effectively negotiate the health care system. "Teach them that if they don't get the support they need in one place, they can reach out to get it elsewhere," he suggests. "Another is to find ways to change the paradigm for these kids' care in an adult setting. How can we adapt strategies that may have worked well for children and make those more adaptable to adult needs?"

As people living with SCD age, the disease worsens as bodily systems break down. "How do you preempt some of those illnesses?" Hoots asks. "How do you get the person the services, the support, the medical access? How do you support them as they move back and forth between having to access health care and access their everyday life in terms of their work and their other colleagues in a broader way than just their health issues?" (These are just some of the questions the NHLBI is trying to answer.)

Meanwhile, Spates and organizations such as SCDAA continue to push for more public awareness about sickle cell disease because there are many misconceptions about the illness. Spates says the craziest one he's heard is that sickle cell is contagious. It's not. Another misconception is that SCD is an African-American disease. This also isn't true. Although in the United States most people with sickle cell are of African ancestry, or identify as black, people who come from Hispanic, southern European, Middle Eastern, or Asian Indian backgrounds can also suffer from the disease.

"Sickle cell is a global disease," Spates says. "Outside of America, there are people from all corners of the world who get this illness."