The Sickle Cell Cure

Once considered a death sentence, sickle cell disease has had a promising medical breakthrough. ESSENCE reports on the cure researchers have been waiting for, dangerous myths associated with the disease and the risks of just being a carrier

BY ERICKA SÓUTER | PHOTOGRAPHY BY JONATHAN SPRAGUE

Mark, 6, saved the life of his 10-year-old sister, Carol.

Lucky Mulumba was at home, holding her 3-week-old baby girl Carol in her arms when the news came. It was a registered letter from the hospital: Carol has sickle cell disease. "I read it four times," says Mulumba, a U.S. Air Force captain and nurse in Fairfield, California. "I couldn't stop crying." Growing up in Uganda, Mulumba had witnessed many children tormented by the disease. "I thought, *How could God do this?*"

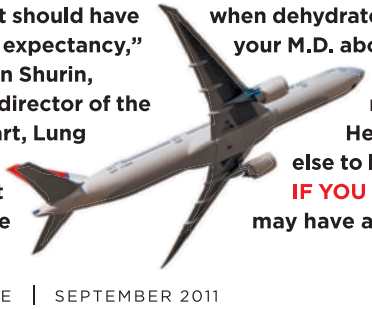
Her fears were understandable. Predominantly affecting Blacks, sickle cell disease causes the body to produce sticky crescent-shaped (rather than circular) blood cells that inhibit the life-sustaining flow of oxygen through one's blood vessels. At one time, children with the disease seldom lived past their teens, and their lives were plagued with chronic pain, deadly infections, kidney disease and even stroke.

It was a reality Lucky and her husband, Abdullah, also a nurse, were not prepared for. They hadn't realized they both carried the trait, which meant their baby had a 25 percent chance of getting the disease.

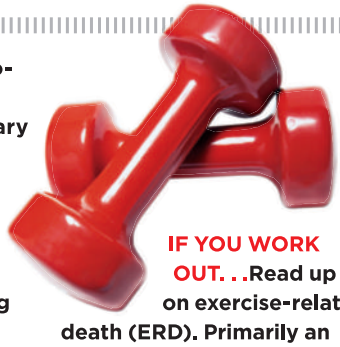
That letter was the beginning of the family's anguish. Whenever Carol got a fever, it resulted in weeklong hospital stays and unrelenting pain. "My head felt like someone kicked it," recalls Carol, now 10. "My stomach was like someone punched it. My foot felt like it was under a rock and my heart felt like it was squeezed." Watching her daughter suffer, Lucky did not plan on more children—but her birth control failed and she learned she was pregnant again when Carol was 2. "I would go ▷

If You Have the Trait...

One out of 12 African-Americans carry the sickle cell trait. There are risks, but don't panic. "People who have the trait should have a normal life expectancy," assures Susan Shurin, M.D., acting director of the National Heart, Lung and Blood Institute. But you may have an increased risk of kidney problems. "Preliminary research shows people on dialysis have a higher incidence of the trait," says Shurin. People with the trait are also more likely to experience blood in the urine when dehydrated. Talk to your M.D. about keeping your kidneys healthy. Here's what else to look for:



IF YOU FLY. . .You may have an increased risk of deep vein thrombosis, a blood clot that can form when you're sedentary for long periods. Spleen infarction, a condition in which oxygen to the spleen is interrupted, is also a risk at 5,000 feet above sea level, according to Michael DeBaun, M.D., pediatrics director at Vanderbilt-Meharry Center of Excellence in Sickle Cell Disease. Come up with a prevention plan with your doctor.

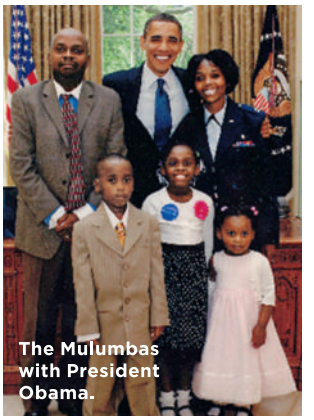


IF YOU WORK OUT. . .Read up on exercise-related death (ERD). Primarily an issue for elite athletes with the trait, the combination of dehydration and exhaustion has resulted in death. Talk to your doctor if you're a hard-core exerciser.

HAIR AND MAKEUP: NELLIE MUGANDA/KENBARBOZA.COM. PLANE: JACK WILD/GETTY IMAGES. DUMBELLS: MARK WEISS/GETTY IMAGES.

in the bathroom and just cry all the time," she says. "Then one day my husband said, 'Just leave everything to God.' And you know, that was the child who saved Carol's life."

By age 6, Carol's health had deteriorated so much doctors said that a bone marrow transplant would be her only hope. Knowing they had run out of treatment options, they moved forward. Air Force doctors worked with civilian M.D.s to make sure Carol got the best care. "A sibling match had the best chance of success," says Michael Grimley, M.D., an associate professor of pediatrics at Cincinnati Children's Hospital Medical Center. It turned out that Carol's brother, Mark, born without the disease, was a near-perfect match. And the Mulumbas had saved 4-year-old Mark's umbilical cord blood which contains stem cells—just in case. "We used chemotherapy to destroy Carol's ability to produce her defective cells," explains Grimley, who performed the transplant at the Methodist Children's Hospital in San Antonio. Then healthy stem cells, harvested from her brother's cord blood and bone marrow, were fed to Carol intravenously. The procedure was not without risk. Though there is an 85 to 90 percent success rate for sibling donations, the surgery proves fatal to some children who fail to tolerate the transplanted blood cells. "Still, we were very positive," says Abdullah. "We had a very strong faith."



The Mulumbas with President Obama.

Within a month, testing showed Carol was cured of sickle cell disease. "She wasn't able to play before, but she is so happy now," says Lucky. The entire family, which now includes daughter Aliah, 3, who does not have sickle cell, visited Uganda last August. And a month earlier on the family's first vacation ever, Carol met her idol, President Barack Obama, thanks to the Make-A-Wish Foundation. As she chatted with the Commander in Chief in the Oval Office, Carol had but one request: "I asked him to take a message to the world—there is a cure for sickle cell," she says. "People need to know. I don't have pain anymore. I started a brand-new life. All kids with sickle cell should be able to start a new life, too."

COULD I HAVE THE TRAIT?

Most states screen newborns right after delivery. Parents, not sure if you carry the trait? Ask your M.D. for a hemoglobin analysis.

The Mulumba family: Abdullah, Carol, Lucky, Mark and Aliah.

4 SICKLE CELL MYTHS

MYTH: Only Black people get sickle cell disease
FACT: Though most prevalent among people of African descent, "It also affects people from India, the Middle East, Greece and southern Italy," explains Althea Grant, Ph.D., chief of epidemiology and surveillance branch, National Center for Birth Defects and Developmental Disabilities.

MYTH: It's a blood disease, so you can catch it.
FACT: Sickle cell is not contagious. "It can't be passed through blood," says Grant. "You get sickle cell by inheriting genes from each parent."

MYTH: Babies born with sickle cell disease usually die in childhood.
FACT: While there was a time when babies with the disease were unlikely to make it past age 14, blood therapies have increased life expectancy by decades.

MYTH: There is no cure for adults with sickle cell.
FACT: The transplant surgery has been less successful in adults who are more likely to reject the donor bone marrow. However, new research from the National Institutes of Health shows promise, so talk to your doctor. "The pool of people who are eligible for a cure has expanded dramatically," says Grant. "That's very exciting."

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