

[back to article](#)



Sickle cell silence

Experts see 'a lack of awareness' of painful condition

*By Naseem Sowti Miller
Staff writer*

Published: Friday, December 26, 2008 at 6:30 a.m.

Three or four times a month, Lynn Davidson gets notification that a newborn has tested positive for sickle cell disease in one of the 23 counties she oversees.

Davidson, a nurse coordinator at the Pediatric Specialty Clinic at Shands Children's Hospital in Gainesville, gets notified because of a state mandate that every child in Florida be tested for the disease and, if positive, must get a follow-up test.

She crosses her fingers every time, hoping she can reach the mother and the baby's pediatrician to immediately put the child on antibiotics and perform the follow-up test. Sometimes the process works smoothly, and sometimes she spends months trying.

The disease is dangerous and can complicate a child's life. There's a month - September - dedicated to sickle cell disease awareness, yet experts say funding and exposure is nowhere close to that for some other childhood diseases, such as cystic fibrosis or muscular dystrophy. Public awareness, said Davidson, even among parents of children with sickle cell disease, is not as great as it should be.

"There's absolutely a lack of awareness," she said. "I'm not sure why there's a lack of awareness. [The disease] is not something that's talked about a lot. It's not advocated a lot. I don't know exactly why that is."

Sickle cell disease is an inherited disorder that affects red blood cells, rendering some of them sickle-shaped. The crescent-shaped red blood cells are not flexible and don't pass through blood vessels as easily as round blood cells.

The disease affects each patient differently and has various degrees of severity. In its worst form, sickle cell puts patients at risk of stroke and causes chronic pain, anemia and a range of other complications that could lead to routine hospitalization. Some patients have to have monthly blood transfusions to lower the risk of stroke.

"The disease has taken a toll on me," said Clifford Kyler, a 49-year-old Ocala resident. "I've had to be hospitalized for blood transfusions. I lost my gall bladder and spleen by 16."

Kyler's recent hospitalizations were in July, August, September and earlier this month, mostly because of episodes of an unpredictable increase in sickle-shaped blood cells

that could clump together and cut off circulation in bones, or various parts of the body, including arms and legs, causing extreme pain.

Many patients have to take medications to manage chronic pain, but then they're at risk of addiction to the medication.

Roughly 75,000 patients are hospitalized for sickle cell disease each year, resulting in direct costs of more than \$475 million, according to a 2006 study published in the journal *Pediatrics*.

The disease brings with it social side effects. Kyler, for instance, has not been able to hold a job because of frequent hospitalizations. His marriage has fallen apart.

"It's been a long journey. And when I look back, there were times that I didn't know I was in this world," he said.

Families of patients with the disease face many issues, said Dr. Julie Panepinto, an associate professor at the Department of Pediatrics of The Children's Research Institute of the Children's Hospital of Wisconsin/Medical College of Milwaukee, Wis.

Panepinto's recent study shows that sickle cell disease reduces children's quality of life, and they usually have learning disabilities and have problems in school.

"They're hit with a lot of things, which impacts their overall well-being," said Panepinto.

In the United States, more than 80,000 people are affected with sickle cell disease. Although the disease can affect anyone, the migration of the sickle cell gene has been such that it predominantly affects blacks.

One in 400 blacks and 1 in 19,000 Hispanics are born with the disease, according to national data.

Yet, compared to cystic fibrosis, which affects 30,000 Americans, sickle cell disease rarely gets as much public exposure or funding.

The National Institute of Health, for instance, allocated almost four times more funding per person affected with cystic fibrosis than individuals affected by sickle cell disease, according to a 2006 study titled "Sickle Cell Disease: A Question of Equity and Quality," published in *Pediatrics*.

"For fiscal year 2003, the Sickle Cell Disease Association of America's revenue was \$458,577, compared with \$152 million for the Cystic Fibrosis Foundation," the study noted.

Funding for the foundations translates into money for research and advocacy.

Davidson works with patients who have sickle cell disease and hemophilia.

"There are massive amounts of resources for my hemophilia patients," said Davidson, the Shands coordinator. "But I don't have that for my sickle cell patients. It breaks my heart to tell these patients that I'm sorry I don't have any resources. I can't even offer them the \$3 parking pass."

Beverly Gilbert, a community outreach coordinator for the Sickle Cell Disease

Association of Marion County, said the biggest request she gets from families is for financial help. "Families usually find themselves in financial strain due to the illness, because either mom or dad can't go to work."

Gilbert helps Davidson reach Marion and Levy county sickle cell patients who need help.

There is speculation as to why sickle cell disease is not better known among the general public and hasn't gained the financial support experts say it needs. One factor many point toward is the role of race.

"There are no similar chronic diseases that primarily affect those from an impoverished, minority background and our findings highlight how this uniquely affects children with sickle cell disease," wrote Panepinto in her study.

Also, researchers of the 2006 study write that, "The question of race has been inextricably linked with SCD since its recognition as a distinct disease. Although it is uncomfortable to contemplate, we must consider the possibility that conscious and unconscious racial bias adversely affects the availability of resources not only for research and delivery of care, but also for the improvement of care."

Despite low funding, the research community has made strides in finding better treatments for the disease. Almost all states test newborns and in most counties, patient advocates educate parents and help them find resources.

Most of the medical costs of children with sickle cell disease are covered by the state's Children's Medical Services and Medicaid.

There's no universal cure for the disease, although a successful bone marrow or cord blood stem cell transplant can potentially cure the disease.

Eleven-year-old Arthur High of Ocala is one of the few in this region to have had a successful bone marrow transplant and has been free of the disease for eight years.

He had a silent stroke when he was 2 years old and couldn't stand or walk for six months, his mother, Margaret, said. Once a bone marrow donor was found for her son, she decided to take the chance and go through with the procedure.

But many parents decide otherwise.

"Once I start educating them about the risks and the chemotherapy that could wipe out [the child's] immune system, they say never mind," said Davidson.

She said most children with sickle cell are on antibiotics and folic acid until age 5.

For those with severe sickle cell disease and pain, a new drug called Hydroxyurea has been helpful, but comes with side effects.

Davidson said she hopes to see a cure for the disease.

"In lieu of that, I'd like to see more support. We need more people like Beverly [Gilbert]. There needs to be support and resources ... there's a huge population and there's a lot of need."

