



Helping Sickle Cell Disease Patients See Hope in an Invisible Illness

Sickle cell disease is the most common inherited blood disorder in the US – but it’s commonly misunderstood as well. Most sickle cell disease patients have no outward symptoms, even as they suffer chronic pain, fatigue and infections. Too often, this invisible illness is minimized, and patients lack the support they need.

Atrium Health offers personalized care to sickle cell disease patients, using cutting-edge treatments and holistic care to ease the pain of this chronic illness.

Learn more about sickle cell disease and how **Atrium Health** helps those who have it.

[Part 1: Game-Changing Advancements in Care](#)

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Atrium Health

Sickle Cell

Part 1:

Game-Changing Advancements in Care

This is part one of a four-part series on Atrium Health's innovative, holistic approach to sickle cell disease care. [Check out the full series here.](#)

Watch VIDEO

About a generation ago, nearly half of the children who had sickle cell disease didn't live past the age of 18. Recent decades, though, have brought advancements in treatments, medications and awareness. People with the disease now live longer and better lives than ever before.

But with the good news comes a new question: How can we continue to improve life for people who will spend decades facing this chronic illness? Atrium Health is at the forefront of tackling this challenge, offering a holistic, patient-centered approach to sickle cell disease care.

What is Sickle Cell Disease, and Who's at Risk?

Sickle cell disease is an inherited disorder that causes red blood cells to be misshapen.

The last few decades have brought drastic improvement to sickle cell disease patients' quality of life. Atrium Health is proud to be at the forefront of these innovations.

Healthy red blood cells are smooth and round, and they're able to carry oxygen throughout the body. But for people with sickle cell disease, red blood cells take on the shape of a C (like a farm tool called a sickle). This leads the red blood cells to stick to each other, forming clumps that block proper blood flow.

When red blood cells can't flow as they should, it can lead to chronic pain, stroke, infection and even organ failure. Sickle-shaped red blood cells also break down faster than healthy cells, causing a shortage of red blood cells known as sickle cell anemia. Sickle cell disease affects approximately 100,000 Americans, with a disproportionate impact upon people of African and Hispanic ancestry are disproportionately affected by the disease.

Suffering from a painful chronic disease disrupts daily life. People with sickle cell disease often require frequent doctor visits, hospitalizations and prescriptions to manage their condition. It can be difficult to go to work or school, and often comes with financial hardship given the level of care that's required. Yet this debilitating disorder often has no outward signs, so the people who deal with it also face stigma about the disease.

"It's hard to have a chronic illness," says [Ifeyinwa Osunkwo, MD, MPH](#) – known as Dr. Ify to her patients – who leads Atrium Health's comprehensive sickle cell disease program at [Levine Cancer Institute](#). "Sickle cell disease will be there all of your life: 40, 50, 60, 70 years. However long you live, you have to work extra hard to take care of yourself on an ongoing, regular basis."

Innovations in Sickle Cell Disease Treatments

During the past few years, Atrium Health has focused on improving outcomes and care for patients with sickle cell disease. The organization offers a comprehensive sickle cell disease clinic that connects patients with medical providers, a psychotherapist, a social worker, a case manager and dedicated nurses who can assist with all aspects of life with the disease.

“We use a multidisciplinary model to determine our patients’ main issues,” says Dr. Ify. “Then we pull from system resources to help address each and every need that the patients have. It’s not just about giving a pill, or a prescription for a medication – it’s a holistic approach to managing the disease.”

The only way to cure sickle cell disease is through a stem cell transplant, where a person with sickle cell disease has their stem cells replaced with healthy cells from a donor. And while this treatment has been around since the 1980s, it hasn’t been widely available. In the past, donors had to be an exact match, meaning they’re the patient’s full sibling, with the same HLA type – a system of genes that one in four siblings share – and without sickle cell disease. These strict criteria meant that only 18 percent of eligible patients could get a transplant.

But through its participation in a national study, Atrium Health has learned that a transplant with a half-matched donor – called a haploidentical transplant – can be effective as well. This could mean that almost every eligible patient could be matched with a donor.

Another common treatment option for patients who have severe or chronic organ complications is a blood transfusion. A donor’s healthy red blood cells are given to the patient, reducing the amount of sickle cells in their blood. And while transfusions are now safer and more effective than ever, they still result in potentially harmful amounts of iron in the blood.

1

Just a generation ago, nearly half of children with sickle cell disease didn’t live past 18.

2

Now, thanks to innovative and holistic care, people with the disease live longer and better lives than ever.

But recently, a safer option has emerged: exchange transfusions. Exchange transfusions draw sickle cell patients’ blood out as they receive non-sickle cell blood, decreasing the amount of sickle cells without creating excess iron. When given monthly, exchange transfusions can significantly reduce the risk of first and second strokes and improve quality of life for children and adults with sickle cell disease.

“The exchange transfusion is a marvelous way of keeping the amount of sickle cells in the blood reduced,” says [Daniel McMahon, MD](#), director of the pediatric sickle cell program at Atrium Health’s [Levine Children’s Hospital](#) (LCH). “It’s relatively quick and easy.”

LCH has partnered with Atrium Health’s Nephrology department to establish a program for regular exchange transfusions. The same principles that have guided nephrologists’ work with kidney disease patients are now helping sickle cell disease patients, too.

By investing in its sickle cell program, Atrium Health has learned more about what patients need and how best to deliver that care to them. The results have been undeniable and immediate – and there’s more to come.

Patient Spotlight:

DeJuan Minnegan

DeJuan Minnegan is only 17, but he's already a veteran of sickle cell disease treatments. He's known since he was a small child that he had the disease, and he's been taking medications for most of his life.

Recently, DeJuan began to receive exchange transfusions at LCH. There is one downside to these transfusions for him - the needles are bigger than regular transfusions, he says - but the positives far outweigh those big needles. His sickle cell levels are lower, and he's even been able to stop taking several of his medications.

DeJuan knows that sickle cell disease impacts his day-to-day life. He knows it causes fatigue that slows him down. He imagines that without the disease, he would've played football like his friends. But overall, he says he's proud to have sickle cell disease. He's proud to manage his health, and he feels the support of his doctors at LCH and his teachers at school.

DeJuan aspires to own his own business one day. In the meantime, the high school junior offers words of support for other young people with sickle cell disease who are just beginning their treatments.

"Don't be nervous. The doctor's got you," DeJuan says. "It's going to help a lot."

His mother, Mary, credits the blood exchange for giving her son a better life, and credits the staff for helping their family navigate this chronic illness.

"There are some people who work for this hospital who have really gone above and beyond to help my family," Mary says. "Things you wouldn't believe, going way past the level of expectations. It's like a family environment."

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Part 2:

Improving Long-Term Care

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Watch **VIDEO**

As healthcare enables people with sickle cell disease to live longer, better lives, the medical community has taken on a new challenge: how to transition patients from pediatric to adult care.

This transition period for young adults between ages 18 and 25 – the years when they need to acquire the knowledge and skills to take greater responsibility for their healthcare – is critical. Yet the number of young adult sickle cell disease patients is beginning to outpace the number of specialists who have the expertise to guide them.

“There is a three-fold increase in mortality rates among young adults with sickle cell disease when they leave pediatrics and enter adult care,” says **Ifeyinwa Osunkwo, MD, MPH** (known as Dr. Ify to her patients), leader of Atrium Health’s comprehensive sickle cell disease program at **Levine Cancer Institute**. “We have to ask: How do we help this niche population?” It’s a big question, and it has elicited many answers that are changing the field, both in new techniques and new technologies.

A New Grant, Funding Big Discoveries

One answer has come through a grant from the Patient-Centered Outcomes Research Institute (PCORI). In September 2017, PCORI awarded a

Atrium Health is making sickle cell disease patients’ transition from pediatric to adult care easier and more effective than ever.

\$9.8 million grant to Atrium Health. Dr. Ify is the principal investigator for the PCORI grant, which funds improvements to the transition of sickle cell disease patients from pediatric to adult care.

One key aspect of these improvements is standardization. All teenagers require the same six steps in their treatment, so Atrium Health has created a continuity of care by continuing those same six steps during their transition into adult care.

“We are at the forefront a groundbreaking intervention for our sickle cell disease patients,” Dr. Ify says. “With this program, patients who are 18 or 19 won’t feel lost. They’ll remain in care. They’ll remain with a provider who knows sickle cell disease, who cares for them and who works with them to ensure that their lives don’t become fragmented.”

Engaging Research and Innovation to Redefine Care

Atrium Health is also participating and engaging in clinical research and clinical trials in the treatment of sickle cell disease. Part of that research involves the development of an app specifically for sickle cell disease patients. The idea is that patients will use the app daily to track pain levels, water intake and medication administrations. By logging these reports, patients and their providers will hopefully be able to identify trends that lead to medical crisis and help prevent those in the future.

As new developments in sickle cell disease treatments continue to improve patient outcomes, the need arises for the medical community to continually raise the bar to keep up with those outcomes – and aim for even better ones.

“One of our biggest goals is to help people with sickle cell disease live longer,” says **Daniel McMahon, MD**, director of the pediatric sickle cell disease program at Atrium Health’s **Levine Children’s Hospital**. “But it’s also to help them become healthy adults with a good quality of life.”

Teammate Spotlight:

Shirley Miller

When Shirley Miller was diagnosed with sickle cell disease at 3 years old, she was told that she probably wouldn't live to be 21. Now in her 60s, Miller is not only maintaining good health, but she's dedicated her career to helping others do the same. She works as a project manager for Atrium Health's sickle cell disease program.

"When I was born, no one really knew a lot about sickle cell. It was a lot of trial and error trying to figure out what was going to help me," Miller says. "I spent a lot of time in and out of hospitals, being stuck with a lot of needles and in a lot of pain. It took a while for my family to even figure out what it was that I had and how to manage the disease."

After beating the odds, Miller decided to become a community advocate for sickle cell disease. While speaking at a national association conference in Dallas, TX, she met Dr. Ify.

Miller was drawn to Dr. Ify's work in transitioning pediatric patients to adult care, assistance that she lacked in her own journey with the disease. After their initial meeting, Dr. Ify offered Miller a role at Atrium Health in Charlotte. Miller is now able to offer the support and information to younger patients that she wishes she had as a pediatric patient.

"Sometimes it's an eye-opener when people meet me because they say, 'You're that old and you're doing well?' And I have to tell them, 'Look, it works. If you do what your care team tells you to do, you can live a long life with sickle cell,'" Miller says.

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Part 3:

Innovation – and Proven – Therapies

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Watch **VIDEO**

The symptoms of sickle cell disease – pain, fatigue and the risk of stroke and infection – can become debilitating to those who face it. As a leader in the treatment and research of sickle cell disease, Atrium Health strives to provide patients with new and alternative sources of therapy to manage their condition.

Managing the Pain of Sickle Cell Disease

Because of the chronic pain that comes with sickle cell disease, long-term use of powerful pain medications, like narcotics, is common. This, however, poses the risk of patients' dependency on pain medications, in which the solution to one problem becomes the cause of another.

The dependence can create continued pain, as the body's tolerance to medication builds. Additionally, withdrawal symptoms these patients may face can be so severe that it becomes

As experts learn more about sickle cell disease, they've found new therapies – including transplants and medications – that can help patients cope with symptoms of the chronic illness.

nearly impossible to maintain a job, attend classes or fulfill the obligations of daily living.

Now, Atrium Health is working with a different drug, buprenorphine, that's helping these patients. The drug has typically been used to treat opioid addiction, but experts have found that it's an effective alternative to a steady pain medication regimen.

Ifeyinwa Osunkwo, MD, MPH, is the first physician at Atrium Health to make buprenorphine a successful treatment option for sickle cell patients. Dr. Ify, as she prefers to be called, is a hematology specialist with Atrium Health's **Levine Cancer Institute** (LCI). She spent months getting trained on buprenorphine from **Stephen Wyatt, DO**, medical director of addiction medicine at Atrium Health, and learning how it could benefit her patients. Buprenorphine helps ease withdrawal symptoms while also treating the pain.

"We have more than 55 patients that we've taken care of with this treatment," Dr. Ify says. "We've gotten them off narcotics – they're out of the hospital, they're getting jobs, starting new careers. It's been amazing."

While buprenorphine doesn't work for all sickle cell patients, it's helped enough to motivate Dr. Ify and her team to share their outcomes with the medical community.

"Atrium Health is leading the charge with this treatment," says **Padmaja Veeramreddy, MD**, who works closely with Dr. Ify. "We're writing papers on our findings, and other hospitals are in the beginning stages of looking at buprenorphine for sickle cell disease treatment. They're looking at Atrium Health as their guide."



For decades, not much changed in sickle cell disease treatment. But now, a surge of innovation is providing patients with new and more effective treatment options – and giving them a greater quality of life.

At Last, a New Sickle Cell Medication

Almost two decades have passed since a new drug was approved for the treatment of sickle cell disease. At last, a new drug arrived last year: Endari.

This medication reduces the inflammation and stickiness that causes red blood cells to sickle. Patients who take Endari experience a 30 to 40 percent improvement in symptoms compared to those who don't take the drug. The result is fewer hospitalizations and pain crises.

Dr. Ify was part of the investigative team that conducted the clinical trial that led to Endari's approval. "I'm so glad it finally got approved. It has a lot of potential," she says. "I'm just so glad that we have another option to give patients."

Now, LCI is leading the charge on increasing sickle cell patients' access to medication. All of Atrium Health's sickle cell disease patients are enrolled in a Patient Management program, which gives them access to Specialty Pharmacy Services. This is a new integrated pharmacy model that works with sickle cell patients to

inform them of the benefits of medications such as Endari, how they may help them, and to find discounts, coupons and payment options to make prescriptions more affordable.

A New Frontier in Sickle Cell Disease Care

The evolution in the treatment of sickle cell disease care has picked up speed in recent years. With new transplant options, a new medication, as well as a new use of an existing medication, providers are discovering new ways to manage patients' symptoms. And as new therapies emerge, Atrium Health is working to make them more accessible and affordable for those who need them, allowing more people who live with sickle cell disease to live with a higher quality of life.

Patient Spotlight:

Evelyn Wilson

When she was 18 years old, Evelyn Wilson went to the emergency room with crisis-level pain. It was then that she learned what was behind the discomfort: sickle cell disease. Though she'd had symptoms of sickle cell since she was 6, it took more than a decade for her to receive a diagnosis.

That day began a cycle of pain treatments, including visits to the emergency room and hospital stays that lasted as long as three weeks. Over time, her body built up a tolerance to the pain medications, requiring stronger doses and additional prescriptions to compensate.

After years of being seen by another hematologist, Evelyn was referred to Dr. Ify to help her cope with the side effects of her sickle cell disease treatments. Dr. Ify suggested trying buprenorphine for her pain.

The medication began a new chapter for Evelyn. She takes buprenorphine twice a day, and since she began taking it, she hasn't had a pain crisis or been to the emergency room.

"The medication actually makes me feel up and lively, with no pain," Evelyn says. "So, I'll put it this way: I smile more. My family enjoys me more. This is really helping."

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Part 4:

An Integrated Pharmacy Model

Watch **VIDEO**

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Atrium Health's Specialty Pharmacy Service gives patients more information, faster service and better outcomes.

The program, known as Specialty Pharmacy Service, has introduced patients to a new integrated model that brings doctors, nurses and pharmacists together. Information is easily and quickly shared, savings opportunities found, and prescriptions fulfilled efficiently. For sickle cell disease patients who face chronic pain, the saved time is invaluable.

"It's a holistic experience," Robicsek says. "We want to erase all barriers to care, so that our patients can focus on what they should - their health, their medications and their treatment options."

The Specialty Pharmacy Service serves a growing number of the region's sickle cell disease patients. The program has already created positive outcomes: Patients begin their medications more quickly, find it easier to stay on them and are more compliant with instructions.

New treatment options. Potential side effects. Questions about affordability. After a patient with sickle cell disease leaves the doctor's office and heads to the pharmacy, new questions about prescriptions often arise.

"There can be so much confusion for our patients as to where to turn or whom to trust," says John Robicsek, Atrium Health's assistant vice president of program development for pharmacy services.

Atrium Health has designed a comprehensive support service for specialty medication management, offering big benefits for sickle cell patients dealing with chronic pain and, often, expensive prescriptions and fragmented pharmacy experiences.

The Right Medication at an Affordable Cost

When Endari earned FDA approval last year, it was the first drug approved for sickle cell disease in about 20 years. It offers patients a new option that, for many, reduces symptoms to never-before-seen levels. Yet, like many new specialty medications, it comes with a high price tag. For patients who face chronic pain that often interferes with daily schedules, the out-of-pocket cost of sickle cell disease medicine can become a major burden, if not an outright barrier to care.

When doctors, nurses and pharmacists work as a team, patients get the best care even faster.

For people dealing with the chronic pain of sickle cell disease, that's welcome news.

The goal of the Specialty Pharmacy Service is not only to inform patients about the newest drugs and their benefits, but to work on behalf of patients to make drugs like Endari more affordable. Team members research copay assistance and payment options that are available, based on each patient's eligibility. They explore all options to reach the lowest possible out-of-pocket cost to the patient. This process allows doctors, nurses and patients to focus more on managing the disease while pharmacy experts coordinate medication processing and affordability.

"What's most important about having an internalized Specialty Pharmacy Service is the ability to have our pharmacists and providers work together to identify patients who could benefit from new treatment options," says Robicsek. "The pharmacy team works to make medications affordable and tolerable, helping patients achieve the outcomes they expect and deserve."

With the Specialty Pharmacy Service, Atrium Health helps secure copay assistance that often exceeds the industry average.

Getting Medications to Patients When They Need It

Sickle cell patients are at risk for pain crises, meaning it's critical for them to maintain the treatment plan laid out by their providers. Atrium Health's Specialty Pharmacy Service works diligently to deliver medications when needed by patients in ways that most outside pharmacies can't replicate. Patients who use outside pharmacies often experience a greater delay in starting treatment.

"Having an internal pharmacy that works within our health system, we are able to deliver a more streamlined and transparent and accountable experience for our patients," says Robicsek.

The Specialty Pharmacy Service allows the pharmacist to work within a patient's health record, greatly reducing the back-and-forth calls between pharmacists, doctors and patients that can delay starting therapy. Pharmacists are also available around the clock, every day of the week, to answer patients' questions.

The goal is to provide holistic care for those who deal with sickle cell disease. Whether they're children or adults, needing a transplant or wanting to learn about new drugs, Atrium Health strives to create an environment where patients feel informed, supported and healthy.

Teammate Spotlight:

LaToya Washington

As a technician with Atrium Health's Specialty Pharmacy Service, LaToya Washington serves as a liaison between the physician, patient and pharmacy. Washington and her team work to remove barriers that stand between a patient and their prescription.

For example, if a prescription requires prior authorization from the patient's insurance, or if the patient needs financial assistance, she works with the team to resolve those issues. When the order is ready to be filled, her department coordinates fulfillment - as well as patient pick up and free delivery to the patient's home, removing transportation barriers.

It's work that's needed for sickle cell disease patients who often face medical crises, Washington says. While new sickle cell disease drugs are on the market and work well for patients, they can be too expensive for many. Washington works with patients to reduce their out-of-pocket expenses as much as possible.

"If a patient didn't have Atrium Health's Specialty Pharmacy Service, we believe their ability to start and continue their prescribed therapy would be greatly reduced," says Washington. "With us, we delve in a little bit more. We seek other avenues, reaching out to the doctors' offices, examining all options. We want patients to focus on getting better and not having that financial burden as well."

And the work that Washington and her team do has already proven to make a difference in patients' lives. With the unique integrated pharmacy model, patients are more likely to be consistent with their medication - and have better health outcomes.

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