ADULT SICKLE CELL Program

ANNUAL REPORT 2017 2018

The **University of Arkansas for Medical Sciences Division of Hematology and Oncology in the Department of Internal Medicine** has partnered with the **Center for Distance Health** to create a statewide system of support for patients with sickle cell disease, and for the physicians who care for them. As UAMS improves and standardizes the treatment of patients with sickle cell disease through the **Adult Sickle Cell Clinical Program,** patients throughout the state with SCD will experience an improved wellbeing.

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From the Director

I feel like I am coming back home.

Recently, I returned to the UAMS Adult Sickle Cell Clinical Program to serve as its interim director as a search is being conducted to find a new, permanent director.

Robin Devan, M.D., and I worked side by side for several months in 2013 to establish the sickle cell program in advance of its public announcement in January 2014. It has been a tremendous success in providing care and guidance to patients with sickle cell anemia in Arkansas and also as an educational tool to providers and fellows.

The program now reaches 240 patients, roughly a third of the sickle cell patients statewide.

The program has helped many people and we are reaching out to more patients every day. With Megan Davis, M.D., and Pooja Motwani, M.D., taking over in 2016, the program after us has remained vigorous, strong and badly needed. The Adult Sickle Cell Clinical Program not only has survived but thrived.

Because the program is a robust and successful one, we're confident it will attract a well-qualified permanent director to lead it, and I am more than happy to help during the transition.

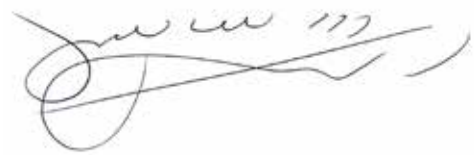
From the inception of this program, Robin Devan and I wanted it to be a repository of clinical data as well as biological data. We worked together to see that happen and collected samples for the disease registry and database. About 90 percent of the program's patients have participated, and the registry is a goldmine waiting for an enterprising researcher to dig into. For someone who is passionate about this field, it continues to develop and grow as a valuable asset for clinical research. The foundations are there on which to build something significant in research.

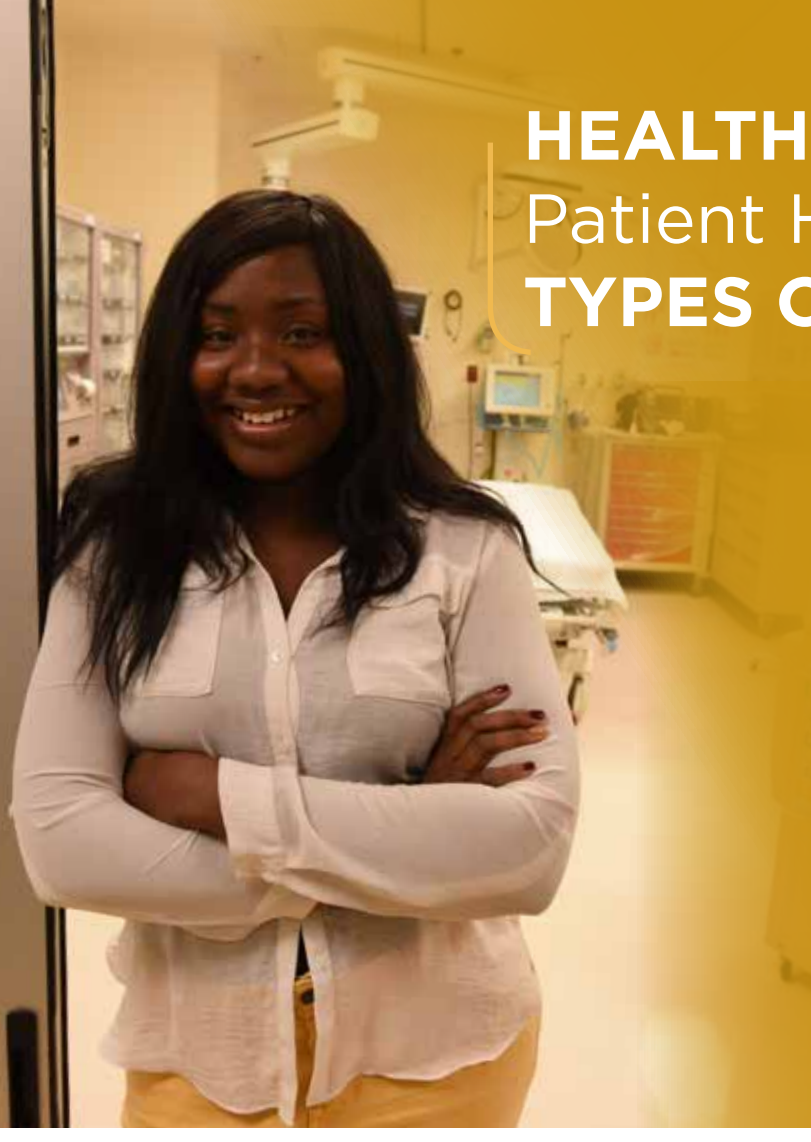
Educating patients, providers and the general public about sickle cell disease remains a core mission of the program, too. Within Arkansas' chiefly rural population, opportunity exists for creative use of distance education on sickle cell disease.

The program's annual symposium has contributed to that educational mission, but continuing medical education is another area for any leader who aspires to make a difference in the community with providers. He or she can use these channels and tools to make sure the program is fulfilling its goals.

The Adult Sickle Cell Clinical Program is meant to be a hub for patients to use one or two times annually not just for health care but to design a care plan for them to follow. That care plan is something patients can take back to their primary care physicians to be implemented locally. With this in mind, we feel the program really is part of a larger, statewide network in which primary care physicians play a central part.

I remain just as enthusiastic about the program now as I did in 2014. Its mission is being fulfilled, and I am confident a new director soon will begin to build on its accomplishments





HEALTH CRISIS Teaches Patient How to Distinguish **TYPES OF PAIN**

By Ben Boulden

Unfortunately for **Joanna Creal-Ward, 29,** pain is familiar enough as she has coped with sickle cell disease throughout her life. But during a pain crisis in March 2017, that familiarity led her to misinterpret the warning signs of another more serious condition.

She went to the Emergency Department at UAMS with the assumption she just would need some pain medication and intravenous fluids to feel better. That's often the type of care she receives at the UAMS Adult Sickle Cell Clinical Program where she has been a patient since 2014.

Fortunately for her, the medical staff decided to take a closer look. They had her undergo a CT scan that revealed pulmonary embolisms —blood clots blocking an artery in the lung that can reduce the oxygen a patient is able to absorb while breathing.

"It's hard to distinguish between the sickle cell pain and the embolism," Creal-Ward said. "It's challenging even for some

of the professionals. They had to do two scans before they saw it. You have to know your body and how your body reacts to different things. I could identify the cause of the pain every time before, but this one time I was wrong. It's hard."

Creal-Ward was admitted as an inpatient to the hospital and expected to be discharged in three days, Creal-Ward said. However, her condition worsened and three days turned into two weeks and a transfer to the intensive care unit.

She learned of her move to the ICU when she awoke there and also discovered a breathing tube had been inserted to allow a ventilator to assist in her breathing. Her physicians and nurses there informed her that her blood oxygenation was at 40 percent of its expected, healthy level.

Creal-Ward said she floated in and out of consciousness while in the ICU. One time on awakening, she discovered her wrists had been restrained because she had been trying to remove the breathing tube while in a semi-conscious state.

"I just started crying," Creal-Ward said. "I could feel the tears flowing down. They gave me something to write with. The first thing I wrote was, 'Where is J?' I have a nine-year-old son, Jadyn. I couldn't write it out though so I just kept writing the letter 'J' over and over again."

Her parents were there and informed her Jadyn was with his paternal grandmother and going to school.

"I wrote 'Pain' and 'Hurt' but they told me I couldn't have the pain meds because it made my blood pressure drop," she said. "So, I told them 'sleep' as in just give me sleep meds so I can sleep. The tube was hurting, and I just wanted to sleep."

Eventually, Creal-Ward recovered as she regained lung function, but her health required her to leave her job as a middle school teacher in Pine Bluff where she had taught for three years.

In late February, she had another pain crisis. Megan Davis, M.D., former co-director of the UAMS Adult Sickle Cell Clinical Program, treated her but also sent her to get a CT just to make sure she wasn't experiencing pulmonary embolisms again.

"For me to get older and for it to get worse with longer stays in the hospital is very frustrating," Creal-Ward said. "I went to school to develop professionally and now I have been sitting out two years. I'm paying for that education, and I want to put it to use."

> "If it feels different from when you usually have one, then you need to get it checked,"

Creal-Ward has a master's degree in counseling from the University of Arkansas at Little Rock.

She admits that sometimes the pain crises are brought on by overdoing. When her energy is good and she's feeling healthy, Creal-Ward wants to get things done and be active.

Her mother, Renisha Ward, reminds her to pace herself. Creal-Ward said she is grateful for that support and the way her mother acts as an intermediary for her with health care providers when she is incapacitated like she was in 2017. Creal-Ward, who now lives in Bryant, also sometimes stays with her mother in

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Patient Makes **'SMOOTH TRANSITION'** into **ADULT CLINICAL CARE**

By Ben Boulden

Turning 21, crossing that last milestone into being legally and fully considered an adult, brought Xavius Hymes a change in his health care that might have worried someone else.

Moving from the sickle disease treatment program at Arkansas Children's Hospital (ACH) to the UAMS Adult Sickle Cell Clinical Program when he came of age didn't concern him much though.

"It was a pretty smooth transition, nothing too major," Hymes, now 22, said. "Both staff are compassionate and really care. I was impressed."

Patients are treated at ACH until age 21. On turning 21, they are transitioned to an adult health care provider. UAMS works with ACH to ensure that transition to health care is an organized process. Hymes said his interactions with the social worker, Leigh Ann Wilson, who works with the patients at both clinics, was both positive and reassuring.

Hymes was diagnosed at birth with sickle cell disease with an SS hemoglobin type. Both his parents carried the genetic trait for the disease. During the following 21 years, the clinicians at ACH treated him and supervised his care. He said he usually made visits to the clinic every one or two months, but at UAMS he rarely needs more than a six-month checkup.

That's largely because as he's matured into adulthood Hymes has felt more knowledgeable about sickle cell disease and better able to take control of his health.

He said he's diligent about making sure he stays hydrated, key to avoiding a sickle cell disease-related pain crisis, and pays close attention to his nutrition, making sure the food he consumes is healthy.

It's been more than a year since Hymes had a pain crisis, and he in part credits the medicine hydroxyurea for that. Hydroxyurea lowers the chances of the clustering of sickle cells that cause vascular blockages, pain crises and other complications.

> "It was a pretty smooth transition, nothing too major"

"If I feel myself edging toward the point of crisis, I try to get fluids and anything else I need," Hymes said. "That usually helps. Nurse Stella Bowers is good about getting me an appointment in the infusion center and that usually helps me avoid having to go to the Emergency Department."

Bowers is a registered nurse in the UAMS Adult Sickle Cell Clinical Program.

Health care has become such a central focus of his life that Hymes' ambition is to secure admission into a medical college. A pain crisis during his undergraduate years forced him to withdraw from classes for an entire semester, so managing his sickle cell disease is key to maintaining his course of study.

In addition to earning his bachelor's degree in biology from the University of Arkansas at Pine Bluff, Hymes is taking classes in Houston so he can score well on the MCAT examine to help him to get into medical school.

Specializing in a medical field may be several years down the road, but he said his current interest is in hematology/oncology or pediatric orthopedic surgery. If he succeeds, then Hymes will complete yet another important transition, the one from patient to provider.

Continued from page 5

Pine Bluff when she has a pain crisis.

"She's my advocate, very much so," Creal-Ward said. The sickle cell program's nurses and physicians also give her supportive reminders and advice so she can stay on track with her care.

To distinguish between pain brought on directly by sickle cell disease and pain from other causes, Creal-Ward advises other sickle cell patients to compare each experience to their past experiences of pain crises.

"If it feels different from when you usually have one, then you need to get it checked," she said. "You also need to evaluate what you have been doing. Have I been hydrating and eating right, over exerting or under a lot of stress? You have to know your triggers."

Adult Sickle Cell Multidisciplinary Clinic

Photo Caption

The UAMS Adult Sickle Cell Clinical Program is a multidisciplinary clinic for adult patients with sickle cell anemia from all over Arkansas. Patients are seen as frequently as needed based on their disease-related complications. This can vary from every month to annually. On the basis of each comprehensive visit, a care plan for that patient is created to serve as a blueprint for the patient's medical care throughout the year and is communicated to each patient's primary care provider.

The Multidisciplinary Clinic of the Adult Sickle Cell Clinical Program at UAMS includes a physician of hematology, Issam Makhoul, M.D.,who specializes in blood disorders and diseases, and treatment of sickle cell disease. Makhoul and a team of hematology physicians coordinate care for patients during their clinic visits.

A nurse practitioner, Collin Montgomery works in collaboration with the team members to deliver care to these patients in the outpatient care setting.

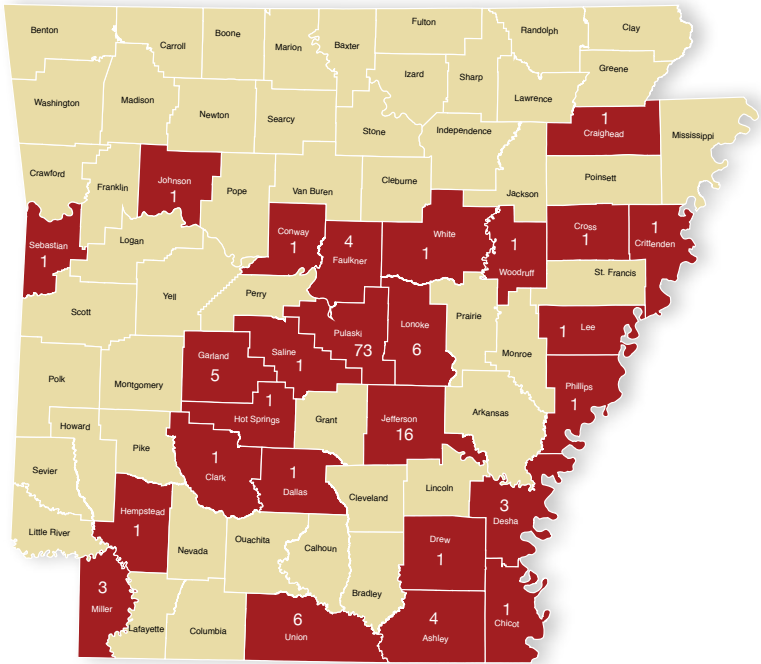
A licensed clinical social worker, Leigh Ann Wilson is in the clinic every week to assist patients and their families regarding social and emotional support, health-related expenses not covered by insurance, transportation costs and employment options. She also is the cornerstone in facilitating successful transition of care from Arkansas Children's Hospital to the adult setting.

A registered nurse, Stella Bowers, assists with providing care to patients in an outpatient setting. She serves as community outreach coordinator connecting the clinic to the community and health care providers around the state. She also serves as the liaison between the call center staff and the sickle cell team, ensuring that the sickle cell team responds to the caller's needs.

A pharmacist, Lindsey Dayer, Pharm.D., assists the providers in delivering medication management through comprehensive medication reconciliation, medication counseling and assessing medication efficacy.

In an effort to provide holistic care, the clinic also assists with referrals for annual ophthalmological assessments and for mental wellness evaluations as needed.

About 900 adult sickle cell patients live in Arkansas. UAMS continues to strive to reach each one. We are committed to providing a better state of health for the sickle cell patients in Arkansas.



INFOGRAPHIC

Program Helps Lake Village Woman **COPE WITH ANXIETY** about Pain

By Ben Boulden

> It feels better to be believed," she said. "I'm glad now that I have someone who believes me instead of someone who tells me I am not in pain or hurting.

For Simone Brown, her fear of pain has become almost as much a presence in her life as the pain from the sickle cell disease she has dealt with since birth. She's not doing it alone, though.

The UAMS Adult Sickle Cell Clinical Program's nurses, physicians and licensed social workers are working with her to reduce her anxiety and the stress it induces, especially because stress itself can contribute to a pain crisis.

Typically, Brown, who lives in Lake Village, experiences sickle cell disease-related pain every day. However, crises of intense pain were almost unknown to her, but in 2006, she had a pain crisis associated with a miscarriage, and another in 2011, when she gave birth to her son.

"When I was younger, I didn't have too many problems with pain, but as I got older it got worse," Brown said. "Three years ago, I had a very bad pain crisis and was hospitalized for an entire week. That was the worst one I have ever had."

The pain was similar to what she experienced before with the two earlier crises but over a longer period of time.

"My whole body hurt with the third one," Brown said. "I was moaning and crying all night. It was horrible, and I was in pain the entire time. It never stopped. They ended up having to give me two pints of blood, too."

Ever since then, Brown has continued to worry about a repeat of that week.

Nevertheless, she hasn't had a pain crisis that severe since, and some of that is attributable to the care she's received from the sickle cell program. Although Brown has been a patient in the program for less than a year, its team, consisting of physicians, an advanced practice nurse, a registered nurse, a social worker and a pharmacist, have helped her manage her pain and her anxiety about it with medications. The program's social worker put Brown in touch with a therapist who can talk to her about her fears and healthy ways of coping with them.

Before coming to UAMS, one source of frustration, stress and anxiety for her were the attitudes she encountered from health care professionals elsewhere who seemed to suspect Brown wanted pain medication for non-clinical reasons or was exaggerating her suffering to get attention. She said they didn't understand sickle cell disease and its symptoms, how real and intense the pain can be.

Her encounters with the program's physicians and staff have been very different and very positive, Brown said.

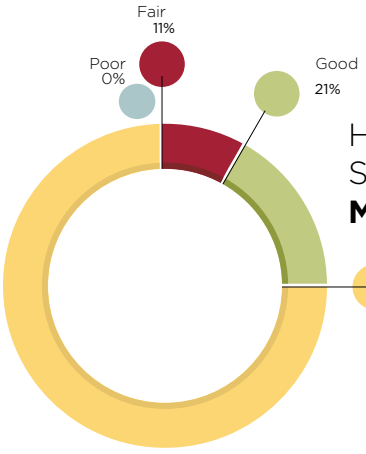
"It feels better to be believed," she said. "I'm glad now that I have someone who believes me instead of someone who tells me I am not in pain or hurting. I hate hurting when someone won't help me to do something about it."

Patient Satisfaction Surveys

In April 2018, the ANGELS Call Center conducted a patient satisfaction survey. The patients surveyed were randomly selected patients of the UAMS Adult Sickle Cell Clinical Program. The survey was specifically about the outpatient clinic experience with the UAMS Adult Sickle Cell Team. About 20 percent of clinic patients received a telephone call from a nurse at the call center and were surveyed with a total of 10 questions. The questions were based solely on the outpatient clinic experience.

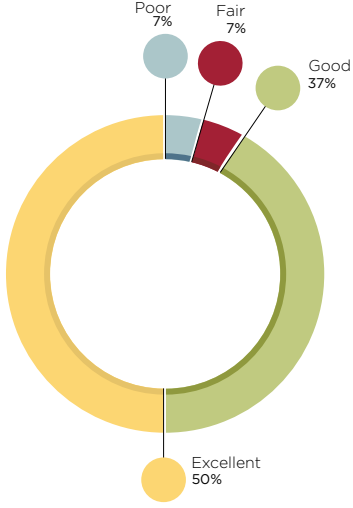
In the past, the survey was written to cover either an inpatient or outpatient experience. This year the questions were modified to better address the outpatient experience and closely resemble the general outpatient clinic survey that UAMS conducts with patients.

All 10 questions received positive responses from an average of 89 percent of those surveyed and many of those being a response of 'excellent.' Areas with the highest positive scores were areas of education: advice on improving health and explaining things in a way that is easy to understand, including medications. In answering questions about these areas, an average of 93 percent of the responses was positive.



HOW DID THE SICKLE CELL TEAM DO AT **MAKING YOU FEEL AT EASE**

HOW DID THE SICKLE CELL TEAM DO AT **LISTENING TO YOUR CONCERNS**

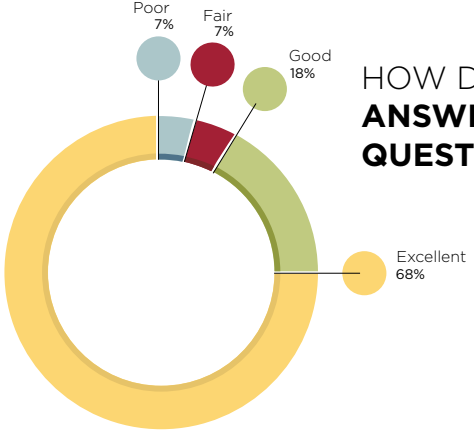


With a goal of improving their quality of life, the Sickle Cell Team realizes that educating patients about their health empowers them to better care for themselves.

Areas with lower total positive scores were areas of communication: listening to concerns, patiently answering questions and showing respect for what patient had to say. In answering questions about these areas, an average of 14 percent of the responses was negative. Although these responses represent a small number of patients, we take this seriously and will be making strides to improve in this area. It is our goal that all of our patients feel that they have had their concerns heard and respected.

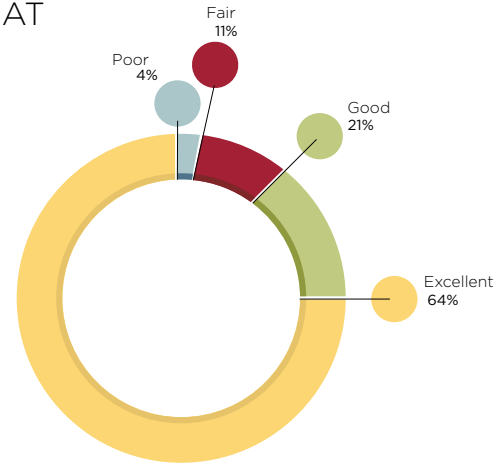
Overall, 86 percent of those surveyed are happy with their care and 90 percent or more would be likely to recommend us to their family and friends. We are very pleased with the overall rate of positive responses. The team will continue to set goals and make efforts toward improvement in the upcoming year.

The graphs below are the results from the Multidisciplinary Sickle Cell Clinic Survey.

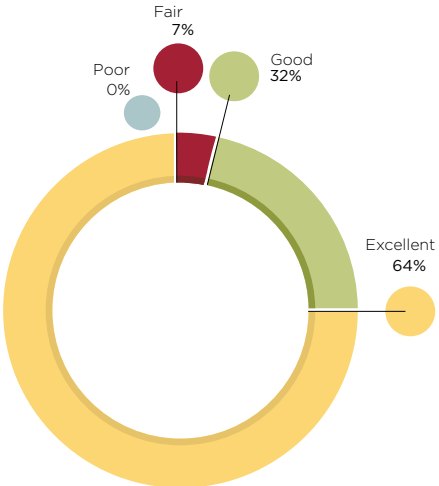


HOW DID THE SICKLE CELL TEAM DO AT **ANSWERING ALL YOUR QUESTIONS PATIENTLY**

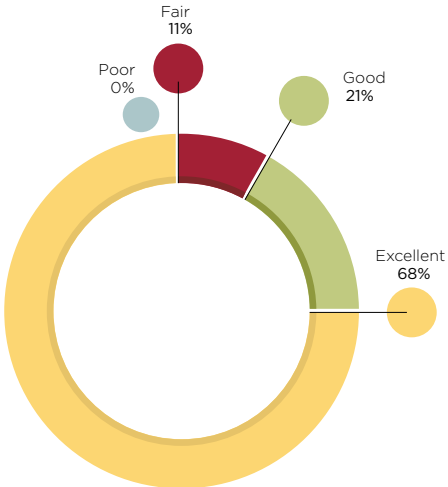
HOW DID THE SICKLE CELL TEAM DO AT **SHOWING RESPECT FOR WHAT YOU HAD TO SAY**



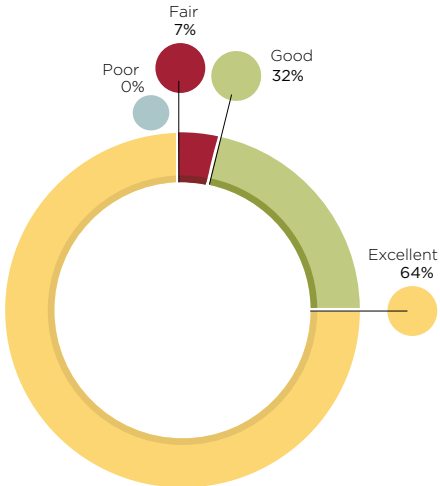
Patient Satisfaction Surveys



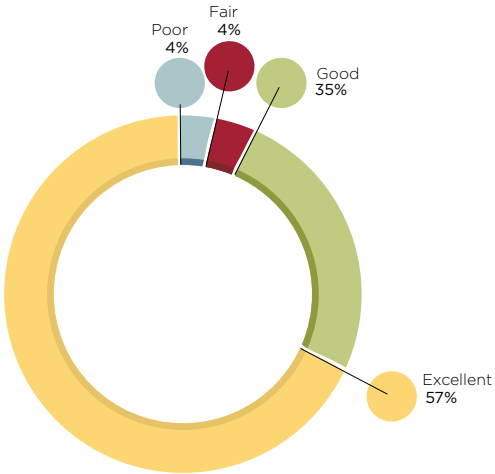
HOW DID THE SICKLE CELL TEAM DO AT **GIVING YOU ADVICE ON IMPROVING YOUR HEALTH**



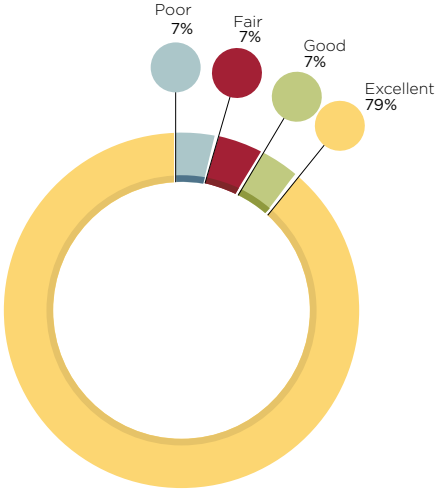
HOW DID THE SICKLE CELL TEAM DO AT **THOROUGHNESS OF THE EXAMINATION**



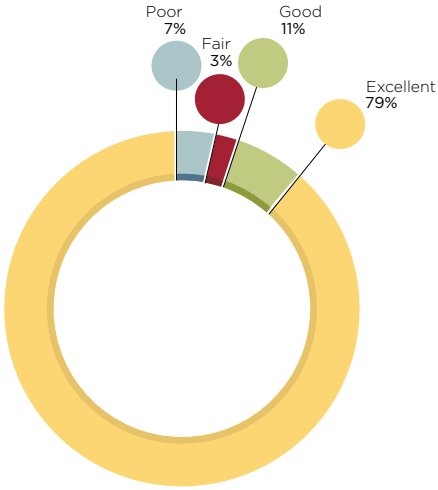
HOW DID THE SICKLE CELL TEAM DO AT **EXPLAINING THINGS IN A WAY THAT WAS EASY TO UNDERSTAND**



HOW DID THE SICKLE CELL TEAM DO AT **EXPLANATION OF YOUR MEDICATION**



OVERALL, HOW HAPPY WERE YOU WITH THE CARE GIVEN TO YOU?



HOW LIKELY ARE YOU TO RECOMMEND THIS CLINIC TO YOUR FAMILY AND FRIENDS?

Call Center

The **24/7 ANGELS Call Center** hosts the **UAMS Sickle Cell hotline** at **1-855-Sic-Cell.**

Patients call in with immediate problems, which are triaged by a registered nurse in the Call Center. The triage nurse may advise the patient to go to the Emergency Department for immediate care, schedule a clinic appointment, outpatient infusion treatment or provide assistance with self-care management at home.

The triage nurse is familiar with sickle cell disease and is equipped with specific triage guidelines of standards of care

disease process. Giving patients direct access to a triage nurse, along with a dedicated Adult Sickle Cell Program Team providing secondary level triage, is reducing admissions and ED visits in the population of patients. The Triage Call Center has been great for the team and the patients. It has provided an option of care and advice for patients other than the ED.

Assisting patients and providers who call is a major part of the call center's function, but at times they have to assist the general public. This year we received several calls from the general public wanting information on our program and asking how they can get involved.

One call comes to mind that involved our outreach component and the call center. A caller stated she had recently been to a community event where she heard information presented on Sickle Cell Disease from the UAMS program nurse. She herself was not directly affected by the disease (she did not have the disease, nor did her family) but was interested in learning more. The call center connected the caller with the RN for the program, and the RN answered all the person's questions and concerns. Increased public awareness for Sickle Cell Disease in adults is critical in changing and improving the stigma associated with the disease.

The call center is a very important component of the Sickle Cell Program and a major player in helping the program to reach its goal of becoming a statewide resource for patients, providers and the general public in the state of Arkansas.

Telehealth Support

24/7 Call Center with Sickle Cell hotline

Staffed by experienced RNs that can offer:

- Patients, families education concerning acute and chronic health problems related to sickle cell disease
- Telephone triage for patients with immediate health concerns
  - Emotional Support, assistance with medication refills
  - Home Care instructions to lessen symptoms, prevent crisis
  - Second level triage (calling Sickle Cell Team Member) before sending patient to ED → giving alternatives to ED visit when appropriate
- Doc to Doc consults - supporting PCPs and ED physicians that are caring for SC patients across the state

Triage outcomes--3 Year Comparison

Caller Request--3 Year Comparison

Education and Outreach

Part of the mission of the Adult Sickle Cell Clinical Program is to ensure adult Arkansans with sickle cell disease receive standardized, comprehensive health care services. To meet that objective, primary care physicians in Arkansas caring for adult patients with sickle cell disease are offered education and support through:

- Collaboration and education regarding best care practices and provider support to keep the patients in their community for care whenever appropriate.

- Telephone consults available 24/7 where the physician can call the Sickle Cell hotline at 1-855-Sic-Cell, and the nurse in the call center will connect them with a physician on the Sickle Cell team. The Call Center nurse will then facilitate any recommendations that a physician needs assistance, hospital transfer, appointment in the sickle cell clinic, or assistance with a copy of the treatment guidelines.

- Evidence-based treatment guidelines on best practices to assist providers in their efforts to care for their patients in their local health care facilities. These treatment guidelines are made available by request and at http://sicklecell.uams.edu/. Treatment guidelines for the management of the perinatal patient with sickle cell disease is available by request on the website, and on the ANGELS guidelines website **http://angelsguidelines.com.**

- Educational presentations, including case presentations, difficult cases and lectures on demand are given to increase awareness regarding best case practices for medical management of these patients. The UAMS

- Center for Distance Health provides the opportunity for participation in Connecting Across Professions (CAP), a statewide means of provider teleconferencing. The CAP teleconferences are given quarterly regarding various aspects of sickle cell disease and treatment. These presentations are available to providers across the state for live viewing and on-demand viewing. Providers can earn CEUs for education provided at these teleconferences.

- The team provides educational lectures to groups at UAMS, including medical residents in emergency and family medicine. The team attends and presents at professional meetings and conferences. The team continues to reach out to health care providers around the state through professional conferences providing disease-specific education and working to identify a core group of providers across the state to engage in collaborative primary care for this population. Outreach is also provided through exhibit booths at statewide conferences.

Educating the Primary Care Physicians during Residency

It is challenging for Arkansas Physicians to provide optimal care to adult sickle cell patients. It is presumed that many adults with Sickle Cell Disease (SCD) in Arkansas don't regularly see a primary care physician. Thus, they are forced to react to their disease crises, instead of focusing on a preventative approach to managing their disease before it becomes critical. This is expensive – not only for the patients, but for the state.

Educating primary care physicians, during their residency, regarding evidence-based standards of care for SCD will promote their level of confidence and comfort in treating adults with this disease when they enter their medical practice. It also is hoped this will result in a reduction in health disparities among Arkansans with SCD because more physicians will be educated to care for adult patients with SCD.

UAMS Adult Sickle Cell Clinical Program is partnering with the UAMS Regional Campuses to educate resident physicians regarding evidence-based standards of care for adult sickle cell disease.

An online curriculum consisting of five sections and 15 modules was developed by the clinical team from the UAMS Adult Sickle Cell Clinical Program, the Arkansas Children's Hospital sickle cell team in collaboration with the UAMS Center for Distance Health and Learn on Demand.

During the five-month course, the residents are asked to complete each subsequent section of modules before a

scheduled monthly didactic video conference. During the video conferences, the presenter (one of our attending physicians that treats adult sickle cell patients), gives case presentations related to the section modules just completed. The residents are encouraged to discuss the cases and to ask questions.

It is our goal that on completion of the following curriculum and didactic case presentations, the resident physicians will be able to recognize the essential components of preventative and acute management of SCD.

The Adult Sickle Cell Clinical Program realizes the **primary care providers of Arkansas need easily accessible resources and support services in order to provide the best standard of care available.**

Education and Outreach

Social Media

In winter 2017, we created a Facebook group for the program. On this social media page, we post clinic information, information on hot topics and links to our patient education modules. We are working to engage more patients to join the group. Patients are informed of the group during clinic and given a flyer on how to find the group on Facebook.

Patient Education Modules

The Sickle Cell team with instructional development specialists at the Center for Distance Health has developed a set of interactive learning modules for our sickle cell patients. These, found at uamspatientslearn.org, provide the patient with information about their disease and appropriate ways to manage their illness. To encourage use of the education modules, patients are assisted in using tablets to access the modules during their clinic wait time. Modules are referenced on the clinic's Facebook page with direct links to make access to the materials user-friendly.

The patient education modules include the following titles:

- Genetics of Sickle Cell Disease
- Nutrition to Fight Sickle Cell Disease
- Respect for Opiates Used to Treat Sickle Cell Disease
- Response to Painful Crises in Sickle Cell Disease
- Medications Used in Sickle Cell Disease
- Organs Affected by Sickle Cell Disease
- Coping with Sickle Cell Disease
- Procedures & Treatments in Sickle Cell Disease
- Pregnancy and Sickle Cell Disease
- Transition from Pediatric to Adult Care with Sickle Cell Disease
- Preventive Health for Sickle Cell Disease

Support Group for Patients and Family

In the fall of 2017, a new platform for the Sickle Cell Support Group began. We collaborated with Sickle Cell Support Services to offer our clinic patients support groups that are hosted in closed groups on Facebook. There are three groups offered by SCSS, Adults Living with Sickle Cell in Arkansas, Caregivers of Sickle Cell Patients in Arkansas, and Kids Living with Sickle Cell in Arkansas ages 7-17. The group members are free to discuss any topics and can share with the group about their life, family and health issues. A patient advocate, LaKisha Johnson of Sickle Cell Support Services, who is not a member of the clinic staff, moderates the support groups and acts as a liaison between the patients and the clinic. The UAMS Sickle Cell Clinical Program encourages participation in the group by distributing flyers in clinic, when attending outreach events and on the program website.

Caregiver's with Adults for Sickle Cell Disease
https://m.facebook.com/groups/2062838997283518

Adults 18 and older in Arkansas living with Sickle Cell Disease
https://m.facebook.com/groups/1952358128319750

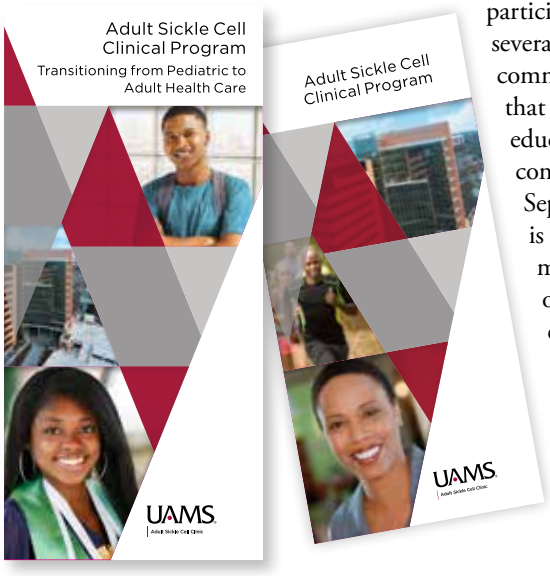
Kids 17 and younger living in Arkansas with Sickle Cell Disease
https://m.facebook.com/groups/296174454199941

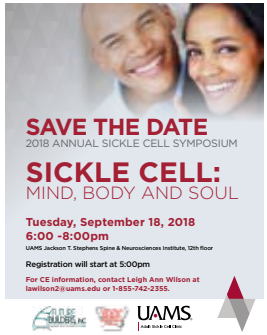
CHARTS

Education and Outreach

Outreach at Health Fairs and Community Events

One of the missions of the UAMS Adult Sickle Cell program is to increase awareness of sickle cell disease with health care providers, patients and community members. The program accomplishes this mission in several ways which include attending health fairs, events at schools, churches and other venues. We continued to partner with Sickle Cell Support Services (SCSS) to provide awareness through the annual Sickle Cell Walk held in April every year. September is National Sickle Cell Awareness Month, and as part of the national campaign, we organized and participated in several media and community events that provide education in the community. September is a special month for our outreach efforts, but we continue to provide awareness throughout the year. Overall for the fiscal year of 2017-2018, we attended numerous community and educational events. See the graph on this page. During these events, we provide information on sickle cell disease and discuss what the program has to offer patients and providers.





In 2017, we hosted our second symposium on the UAMS campus titled: "Stem Cell Transplant as a Cure for Sickle Cell Disease: Who, When and How?" Our partners, Future Builders and Little Rock Black Nurses Association of Arkansas, help to coordinate this event. Our keynote speaker was Kathryn Yarkony, R.N., Ph.D. Transplant Coordinator, at Johns Hopkins University. She provided our patients and local providers with valuable information on stem cell transplants. Pooja Motwani, M.D., associate professor of hematology and co-director of the UAMS Adult Sickle Cell Clinical Program provided information on how UAMS could assist in the stem cell process. UAMS coordinates a patient's care with Johns Hopkins to provide this treatment option, and the university looks forward in the future to providing this treatment directly to patients.

Outreach will continue to be a major component of our program because the more we reach out, the more patients we will be able to assist. In our outreach process, we also are building relationships with physicians throughout the state to be able to provide hometown primary care for patients. We will continue to raise awareness and to provide education to as many patients and providers in Arkansas as we can reach.

Symposium Focuses on BONE MARROW TRANSPLANTS for Sickle Cell

At the end of the Sickle Cell Symposium on Oct. 19 at UAMS, a young woman at a microphone during a question-and-answer session boiled the evening's presentations down to one question: "I haven't heard the word 'cure' used much tonight. Can you call this a cure?"

Pooja Motwani, M.D., said, "Yes, you absolutely can use the word 'cure.' You can."

Motwani makes her presentation at the symposium

One of 59 who attended the symposium, the questioner was specifically asking Motwani about the promise of half-match bone marrow transplants for curing sickle cell disease, the symposium's focus.

Motwani, co-director of UAMS Adult Sickle Cell Clinical Program, was the second of two speakers at the program, and her presentation was titled "Stem Cell Transplant as a Cure for Sickle Cell Disease: Who, When and How?" She followed keynote speaker Kathryn Yarkony, Ph.D., A.P.R.N., who gave an overview of the bone marrow transplant program at Johns Hopkins University in Baltimore where she is the lead transplant coordinator.

Bone marrow transplants are one of the procedures used to restore stem cells. Recent advances in 'half-matched' bone marrow transplantation have allowed for a much broader range of donors, allowing just about any patient to be eligible for the procedure. This means a donor can be found for nearly every patient who needs a bone marrow transplant to be cured, even if the match isn't exactly the same.

"The transplant center at Johns Hopkins has one of the best, if not the best, when we talk about outcomes for patients for

Kathryn Yarkony, Ph.D., A.P.R.N., answers a question from the audience as Pooja Motwani, M.D., listens at the Sickle Cell Symposium.

transplant, pediatric and adult transplants," Motwani said. "They have very little mortality and very few instances of graft-versus-host disease with their current regimen."

Graft-versus-host disease occurs when a recipient's body accepts the new bone marrow but begins attacking the recipient's own cells.

Yarkony said Johns Hopkins has done transplants for 55 patients — two-thirds were sickle cell patients. More than 90 percent of those sickle cell patients have had new bone marrow cells successfully grafted into their bone marrow. Once the new bone marrow cells take hold, she said, the new cells start producing a new immune system and new red blood cells in the patients. These red blood cells are healthy and do not cause sickling.

While the process cannot reverse some of the effects of the disease, it can stop its progression and just as importantly, successful transplant patients no longer experience the pain crises that disrupt their lives, often result in disability and send them into emergency departments and the hospital, Yarkony said.

Both Yarkony and Motwani said there are some risks with bone marrow transplantation. The chemotherapy used prior to implantation to wipe out a patient's bone marrow to make room for the new cells can result in infertility and often produces temporary side effects like hair loss, severe fatigue and nausea.

Obstacles to receiving the treatment include the need for pharmacy coverage, insurance and housing while at Johns Hopkins. Transplant there requires a stay of two to three months and the constant presence of a caregiver.

Clinical social workers are ready to work with transplant patients on finding grants and other sources to help finance the procedure, Yarkony said.

Motwani said the half-match has greatly increased the options for sickle cell patients, but there are 47 clinical trials around the country being done at various centers where stem cell transplants are being used as a strategy to cure sickle cell disease.

Sickle Cell Disease Registry

A Sickle Cell Disease Registry was established to learn more about adults livingwith the disease in Arkansas. Before this,the state did not have a way of tracking patients with sickle cell disease after newborn screening. It is still unclear the exact number of people living in Arkansas and the United States with sickle cell disease. The Arkansas Department of Health has been conducting newborn screeningsof all Arkansas births since 1988, which including newborns withsickle cell disease. From the newborn screenings,we know about 25 babies are born each year in Arkansas with sickle cell disease. Given that those with themost severe form of the disease have a life expectancy in the mid-40s,we canestimate atotal number of people with the disease in Arkansas at 1,300. We also know that about 450-500 of these patients are pediatric patients,which means about 800-850 are adults. Patients are eligible for participation in the registry if they are at least age 18, live in Arkansas and have sickle cell disease. Patients are identified for the registry by the sickle cell team at UAMS. Patients have the ability to participate in the data collection portion, as well as a one-time collection of a blood and urine sample. The blood and urine samples are being collected and stored for future research. Collection for the registry began in March 2015,and as of March 2018,108 patients have consented to participate in the disease registry. Of the 108 patients, 92 patients have provided blood and urine samples. Reports for the data have been completed with all 108 patients and are represented in the following graphs. Participants ofthe Sickle Cell Registry alsoareasked to complete a Quality of Life survey. The Quality of Life (QOL) survey is defined as an overall assessment of a person's well-being, which may include physical, emotional and social components as well as stress level, sexual function and self-perceived health. The QOL helps providers better understand the impact that sickle cell disease is having on a patient's daily life. The QOL ranges from 16 (worst possible) to 112 (best possible). The graph represents the numbers for the QOL data collected in the disease registry.

INFOGRAPHIC

UAMS

Adult Sickle Cell Clinic

To utilize the service of this program
call the 24/7 Call Center
@ 1-855-Sic-Cell (742-2355)
For more information visit
http://sicklecell.uams.edu