University of South Alabama
Comprehensive Sickle Cell Center
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# POETRY COMPETITION

All sickle cell disease clients are asked to submit their favorite, original poem for publication in the September 2013 newsletter.

Your poem should be submitted to Mrs. Brittany Brown at brittanybrown@usouthal.edu in pdf format by July 31, 2013.
The top three poems submitted will receive a prize of $50 for first prize, $35 for second prize, and $25 for third prize.
Only one poem per client can be submitted.
The Sickle Cell Center faculty and staff look forward to sharing in your creative writings.

Johnson Haynes, Jr., MD
Director, USA Sickle Cell Center

Visit the Comprehensive Sickle Cell Center website at:
http://www.usahealthsystem.com/sicklecellcenter

Sickle Cell Today
USA Comprehensive Sickle Cell Center

Volume 10 Issue 1 April 2013

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USA Comprehensive Sickle Cell Center
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Adult Clinic (251) 470-5890
Community-based Program (251) 432-0301

# Back to the Basics: Sickle Cell Disease 101

Ardie Pack-Mabien, CRNP

Sickle cell disease (SCD) is a hereditary blood disorder that affects red blood cells (RBCs) in humans. Hereditary means both parents must have sickle cell trait or another abnormal form of hemoglobin, such as hemoglobin C, for the child to have SCD.

Unlike the normal RBC that resembles a donut, RBCs in SCD are abnormally shaped in the form of a sickle, or crescent shape moon. This change in the shape of the RBCs occurs when oxygen is released from hemoglobin (a protein inside the red blood cell). These sickle shaped RBCs can cause blockages in blood vessels, thus stopping the flow of oxygen to the body's organs (brain, bone, skin, eyes, heart, lungs, kidney, liver, and spleen).

SCD can cause pain and even death from complications such as stroke, lung disease, kidney failure, and infections. Sickle cell anemia, the sickle beta thalassemias, and sickle C disease are the most common forms of SCD. SCD is seen in people of African, Mediterranean, Indian, and Middle Eastern descent. In the United States, SCD is the most common hereditary disease known and is most commonly seen in African Americans. The largest number of non-African Americans affected by SCD in the United States is the Hispanic population.

SCD continues throughout the life span for those affected with the disease and is not contagious. The first three years of life are the most crucial because this is the period of time associated with the highest incidence of mortality. The prognosis for individuals with SCD has improved significantly over the years as a result of mandatory newborn screening with early diagnosis and intervention, preventive medicine practices, and hydroxyurea therapy. Preventive medicine can often delay or slow the progression of complications associated with SCD.

The average life expectancy for individuals with sickle cell anemia is 42 years for males and 48 years for females and for individuals with sickle C disease, 60 years for males and 68 years for females. The oldest living male and female with sickle cell anemia seen at the University of South Alabama Comprehensive Sickle Cell Center (USA-CSCC) is 70 and 65 years of age, respectively. The oldest living male and female with sickle C disease at USA-CSCC is 61 and 76 years of age.

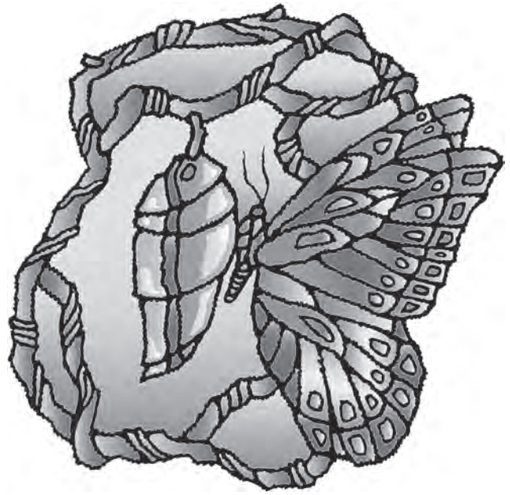
Individuals with SCD should receive regular care from a knowledgeable healthcare provider (hematologist, sickle cell specialist) in combination with their primary care provider (pediatrician, family medicine, or internal medicine healthcare provider). Illnesses affecting the general population also affect those living with SCD such as high blood pressure, diabetes, and cancer. Comprehensive outpatient care can reduce morbidity, lessen the frequency of complications, speed rehabilitation, and ease mental stressors. Preventive medical practices include immunizations, folic acid, penicillin prophylaxis, compliance with diagnostic studies, and an annual vision exam by an ophthalmologist (eye specialist). Individuals with SCD should schedule an appointment with their primary care provider and obtain a referral to see their hematologist or sickle cell specialist!

Contact Information:
USA Pediatric Hematology/Oncology
Pediatric Sickle Cell Center
1504 Springhill Avenue, 5th Floor-Cabana Row
Mobile, Alabama 36604
(251) 405-5147 front desk

USA Adult Sickle Cell Clinic
Mastin Physician Group
2451 Fillingim Street, Suite 102
Mobile, Alabama 36617
(251) 470-5890 front desk

# Checking Your to-do List for Transitioning

Brittany Brown, RN, BSN - PACT Coordinator



Do you know the correct way to transfer from pediatric care to adult services? Do you know what paperwork and insurance information are required for successful transfer to adult services? Listed below is a step-by-step approach outlining the transfer process to adult services at the University of South Alabama Comprehensive Sickle Cell Center. The PACT (Pediatric to Adult Care Transition) program begins preparing the pediatric clients for transitioning to adult services starting at age 13 years. The pediatric physicians continue care through the age of 19. Once the transfer process has been completed, (clients will be notified at their last pediatric clinic visit) the client has a 30 day grace period to contact their pediatric providers for questions, concerns, and medication refills.

Check List:

1. Define a primary care provider (PCP) if one has not already been established. Please confirm that your chosen PCP sees adult patients. Patients are required to maintain a relationship with their PCP for health issues other than sickle cell disease.

2. Determine if your health insurance requires a prior-authorization or referral to see a sickle cell specialist. Your PCP will provide the necessary paperwork. Please request that prior-authorizations and referrals be faxed to the adult sickle cell clinic at 251-470-5895. Prior-authorizations and referrals are needed prior to an adult clinic appointment being made.

3. Medical Records are required prior to your adult sickle cell clinic appointment. As a transferring pediatric patient from the USA system, medical records will be obtained from the USA pediatric sickle cell clinic prior to your adult clinic appointment by the transitioning nurse coordinator (Mrs. Brittany Brown). Please notify the PACT coordinator if your demographic information, including current mailing address and contact numbers, has changed.

Please contact Brittany Brown, R.N. at the USA Comprehensive Sickle Cell Center at 251-470-5893/ 470-5875 for questions or concerns regarding a new patient appointment with our adult sickle cell providers, Dr. Johnson Haynes, Jr. and Ardie Pack-Mabien, C.R.N.P. Please be mindful that appointments are not made, until all required paperwork, as listed above, has been received in our office. New clients will be notified by our office with their new patient appointment date, time, and the clinic location. We look forward to seeing you on the adult side!

*Transition in a word means… Change!*



# THE SOCIAL WORKER'S CORNER

The Knights of Peter Claver, Inc. and Ladies Auxiliary award grants to individuals affected by sickle cell disease (SCD) and to institutions or individuals that provide research in this area. The Sickle Cell Disease Association of America-Mobile Chapter, Inc. was successful for the third consecutive year in securing grant funding for a client with SCD that resides within the nine-county service area.

The most recent recipient for the Knights of Peter Claver National Sickle Cell grant is Johnyell Rodgers of Jackson, Alabama. Mr. Rodgers has experienced many complications related to SCD. This grant will assist Mr. Rodgers in covering many of the costs typically associated with managing this type of illness. The Knights of Peter Claver Ladies Auxiliary, Court 1 provided this award to Mr. Rodgers in the fall of 2012. Alisha Grayson and Dexter Jones were recipients of this award in 2010 and 2011, respectively.

The Sickle Cell Disease Association of America-Mobile Chapter, Inc. and The USA Sickle Cell Center would like to thank the Knights of Peter Claver, Inc. and Ladies Auxiliary for their continued support of the sickle cell community.

If you would like more information in regards to this or any of our community service projects or programs, you may contact us at (251) 432-0301.

Until next time, so long from the social worker's corner.

Adrienne Petite, LBSW
Social Worker Case Manager
SCDAA-MC

# Graduating Class of 2012

It is with great joy and pleasure that the University of South Alabama Comprehensive Sickle Cell Center take this opportunity to congratulate the graduating class of 2012 on a job well done. These individuals have endured medical challenges associated with their disease in conjunction with the day-to-day academic challenges. They have met their challenges with unrelenting hope and determination and are now graduates. The following individuals have successfully completed high school, college, technical school or a career development program. We encourage all of the graduates to continue your education and/or training as you prepare yourself for the ever competitive and demanding job market.

2012 High School Graduates:

Stanley Barnes, Jr.
Kendrae Dennis
Kenisha Dennis
DeJuan Edwards
Javares Gebrehiwet
LaClara Peyton
Reginald Rodgers
Tovah Williams

2012 College and/or Vocational Program Graduates:

Kimberly Burden
Kathryn Davis
Joshua Evans
Essie Jackson
Jeremy McDonald
Hiram Wright


*Congratulations from the USA Comprehensive Sickle Cell Center!*

# USA ANNUAL SICKLE CELL CONFERENCE 2012: A Legacy of Excellence

Johnson Haynes, Jr., MD

Over 70 participants consisting of physicians, physician assistants, nurse practitioners, registered and licensed practical nurses, pharmacists, social workers, and staff attended the 2012 conference. The 2012 conference was the 12th conference conducted by the USA Sickle Cell Center. The conference theme is geared annually to address practical issues in medicine that impact the care of patients affected with sickle cell disease (SCD). Unique to the 2012 conference was the emphasis on a more holistic approach in the care of sickle cell clients, which entailed discussions on psychosocial, ethical and medical issues involved in effective healthcare delivery. In addition, the 2012 conference was expanded from a one day to a two day conference.

The Dr. Cecil L. Parker, Jr., Sickle Cell Disease Distinguished Lectureship was presented by Dr. Robert "Bob" Adams from the Medical University of South Carolina in Charleston. He lectured on how the early identification of children at risk for stroke using the transcranial doppler and implementation of chronic red blood cell transfusions have resulted in a falling rate of childhood stroke in SCD. The 2012 conference featured a second guest lecturer, Dr. Alan Sanders, Director of Ethics at Catholic Health East in Newton Square, Pennsylvania who spoke on "Ethics in pain management, fears, realities and needs." The conference also featured University of South Alabama faculty. Dr. Thomas Butler, Associate Professor, Mitchell Cancer Institute, lectured on "Platelet disorders in sickle cell disease." Dr. Johnson Haynes, Jr., Director, USA Sickle Cell Center, discussed the potential challenges of sickle cell disease in air travel. Dr. Susan Baker, Associate Professor, Department of Obstetrics and Gynecology, addressed issues in pregnancy unique to SCD. Dr. Felicia Wilson, Professor of Pediatrics, discussed newborn screening in sickle cell disease. Dr. Juvonda Hodge, Assistant Professor, Department of Surgery, discussed the clinical indications and special considerations involved in performing cholecystectomy in the sickle cell disease client. Dr. Anthony Martino, Professor and Chair, Department of Neurosurgery, lectured on the pros and cons of encephaloduroarteriosynangiosis (EDAS) in managing moya moya in sickle cell disease. Dr. Elise Labbe-Coldsmith, Professor, Department of Psychology, discussed psychosocial issues commonly seen in individuals affected by sickle cell disease. This year's conference included interesting cases that were presented for discussion by Dr. Zakiya Douglas, Fellow, Pulmonary and Critical Care Medicine; Dr. Sheryl Falkos, Associate Professor of Pediatrics and Dr. Michelle Grier-Hall, Pediatric Medicine, private practice.

Many thanks for the financial support from the USA Medical Center Auxiliary and Center for Healthy Communities. Their support has been vital in keeping the cost of meeting registration affordable and has enabled the Sickle Cell Center to provide affordable continuing education for healthcare providers in the communities we serve.

The next annual conference will be held in the spring of 2014. Congratulations to Mable Barron, LPN, winner of the early bird registration drawing for complimentary admission to the 2014 Annual Regional Sickle Cell Conference. For additional conference information and your chance to win complimentary admission for the 2015 conference, call (251) 470-5893.



# Annual Blood Drive Makes Goal: Youth on the Move and Making Strides in 2012

**Ardie Pack-Mabien, CRNP**

The USA Comprehensive Sickle Cell Center staff, Alpha Phi Alpha Fraternity, Inc, Franklin Primary Health Center, and the Sickle Cell Disease Association of America, Mobile Chapter 2012 Blood Drive was a great success. This partnership began in 2005 and is conducted during the month of September in recognition of National Sickle Cell Awareness Month. The 2012 blood drive was held on Saturday, September 22, 2012 at the Franklin Memorial Complex Mall located at 1303 Martin Luther King Avenue, Mobile, Alabama.

Many lives in the community have been affected by the local blood drive and each year it has grown in participant numbers and success. There was an enormous amount of support from the youth of this community at the 2012 blood drive. Youth supporting the blood drive included University of South Alabama medical and physician assistant students, the Alpha Elites, and local area, middle, high school, and college students. Community youth volunteer efforts included registering donors, serving refreshments, prize distribution, and blood donation. Blood drive sponsors were extremely proud of the efforts put forth by our youth as they make strides toward a more positive future. While the goal of this year's blood drive was to collect 44 units of blood, the goal was exceeded with 49 units collected. Sixty-four people presented as possible blood donors; eleven more than the previous year and seventeen were first time donors. Forty-one participants were able to donate. Four donors gave what was equivalent to two units using the ALYX procedure. Each unit of blood obtained was separated into red cells and plasma potentially effecting the lives of 141 individuals.

For the third consecutive year, the Pacesetter Motorcycle Club set the standard of community involvement with the highest group participation for blood donors; "Job Well Done" goes out to each and every volunteer at the 2012 blood drive. Thank you for your dedication and continued support and for giving the "Gift of Life" through blood donation. The life you save may be yours, your family, or friends. We hope to see you and your organization at the 2013 blood drive.

The 2013 Blood Drive is tentatively scheduled for Saturday, September 21, 2013 at Franklin Primary Health Center located at 1303 MLK Drive, Mobile, Alabama.


L to R: Shernell Mabien, Austin McGrew-Haynes, and Brittany Brown, RN

# From the Director's Desk:
## *Who Would Have Thought?*

I am a third generation graduate of Meharry Medical College. After graduating number one in my class in 1980, I began an emergency medicine residency at Beth Israel Hospital, which is a major teaching hospital of Harvard University. While at Harvard, I quickly gained the respect of the faculty and of my colleagues as being a future leader in Emergency Medicine. The division director, Dr. Diggs, and I had just recently returned from a national meeting where I presented some of our work on the emergency room management of asthma and a new technique to stabilize the chest wall following blunt trauma. This work was receiving much attention from the emergency medicine community. Because of these scholarly activities, I was being heavily recruited to join the faculty at Baylor College of Medicine. After a series of interviews, I decided to join the Baylor faculty as an assistant professor of medicine. There I continued my research, teaching, and clinical activities. I was referred to by many as a "triple threat." This once was the expected but because of changing economics in medicine, very few physicians in academic medicine were able to achieve this. This all served me well. I rose quickly through the ranks at Baylor to professor of medicine, which led to the next phase of my professional life. It was now time for me to begin looking at division director jobs in emergency medicine. As I combed through over ten potential positions listed in various journals, I finally saw a position available at what I thought was the perfect place, Beth Israel. The next day, as fate would have it, Dr. Diggs called and asked me if I would consider moving back to Boston as the Director of Emergency Medicine. He stated he was retiring for medical reasons and thought I was the perfect person for the job. I did not want to appear too anxious so I told him I would love to come for an interview but if all else failed it would be good just to see him again. As I hung up the phone I was so excited and yes, I wanted that opportunity. The initial interview was scheduled for four weeks.

Those four weeks were the longest four weeks of my life. Finally, the day came and I was off to Boston. As I boarded the plane, I noticed a pain in my lower back and right arm. This was a pain that I was quite familiar with in my younger years but since I had been on a new medication, Hydroxyurea, I had not had any pain for over three years. Today I was not about to let this pain impair, in any way, my chances of becoming the new director of emergency medicine at Beth Israel. So I took some pain medicine and felt much better. I immediately forgot about the pain as my mind raced in delightful thoughts about my bright future at Beth Israel.

When the plane landed, Dr. Diggs met me at the baggage claim area. We shook hands and proceeded to retrieve my luggage. I was feeling so many things from nervousness to excitement to an occasional twinge of pain. The only thing I could figure was that some how the excitement was triggering a mild pain crisis. I was determined it would not get the best of me. Dr. Diggs went on to tell me he had been diagnosed with ulcerative colitis about two years ago and would need surgery soon. Because of this and rapidly approaching 70, he thought it was a good time to retire. I in one breath told him, "70 isn't old and after you recuperate from your surgery you will feel differently." He said, "I have run my race and it's time I pass the baton to you. Beth Israel needs you." I was flattered but with a sense of sadness. Dr. Diggs was the doctor who had shaped my career; the physician I had always wanted to emulate. It just didn't seem right he was going to retire and I would possibly take his place. All I could think of was what big shoes I would have to fill. As we drove to the hotel, he said dinner was at 7 p.m. That gave me about three hours to unwind and let what was happening sink in.

At dinner there was the dean of the medical school, the chairman of medicine, Dr. Diggs and myself. It was flattering to be in the company of such esteemed members of the Harvard faculty, particularly on the first interview. It was an amazing evening. It was clear they truly wanted me to take the position. This was capped off with a wonderful merlot and toast to the success of the program. The evening ended and Dr. Diggs took me back to the hotel. The next day was to be filled with interviews by the faculty and the dean, to be followed by a tour of the facilities. With all this in mind I thought I had better get some rest. That night was a long, restless night. All I could do was think about the next day. When I arose the following morning, I felt as if I had been in a wrestling match. After a hot shower, I felt better and got ready for the interviews.

My first interview was with a young, energetic, associate professor in the emergency department who felt her research was of primary importance at this time in her career. Clearly, she would be a major player in maintaining a strong academic program. Following our interview she introduced me to all of the residents in the program. They reminded me of myself some years ago and were obviously very bright. From there I met with one of the more senior staff. He looked me over from top to bottom, as if to say, I was too young. As the interview went on, it became clear he loved this place and was an excellent teacher. I left him thinking, if for no other reason than his love for the place; we would be able to work things out. Following several more interviews with faculty, the day ended with Dr. Diggs. He asked what did I think? I told him I thought this was a great place to be a part of and would love to take the job. I knew ultimately it was up to the dean of the medical school but that Dr. Diggs could influence his decision. Dr. Diggs and I stopped and had a quiet dinner. My mind raced with the thoughts of becoming the new director. I am sure I seemed distant during dinner but I knew he understood how I felt. After all he was also a little distant and consumed with the changes about to occur in his life as well. We returned to the hotel. I had a late morning flight back home the following day. I told Dr. Diggs not to worry and I would just catch a cab to the airport in the morning. He said okay and to look to hear from the dean in about two weeks. We said good night and I went to my room.

That night about 10 p.m. my legs and back started hurting. I took something for pain but did not get any relief. Three hours later the pain was some of the worst I had ever experienced. I called the front desk and told them to have me a cab waiting because I would need to go to the emergency room at Beth Israel. Looking a bit disheveled, I came down obviously in pain and was carried to the emergency room. When I arrived at the emergency room I told the receptionist I had sickle cell disease and was having a pain crisis. She took all the necessary information for registration and told me to have a seat and that the nurse would be with me shortly. After about an hour the nurse finally took my vitals and put me in an exam room. As I lay there writhing in pain the nurse was able to place an IV after four attempts. The pain continued to escalate. Seemingly, everyone was moving so slowly. Finally, one of the residents I had met during my interviews came in to evaluate me. When he finished, he stepped out of the room and told the nurse to give me 25mg of Demerol. I heard him say, "that patient is not really hurting that bad." He obviously didn't recognize who I was. The nurse brought in and administered the Demerol as instructed. I must have dozed for about ten minutes but awoke in extreme pain. I asked the nurse for more pain medicine. When she told this to the resident, he replied, "that patient was asleep just five minutes ago; can't be hurting so soon." Reluctantly, he gave me an additional dose of Demerol and finally the pain was significantly relieved. Two hours later I received more pain medicine and felt much better. This along with the IV fluids had me feeling well enough to return to the hotel. This was an unusual experience for me. On one hand, at home everyone knew me and treated me aggressively and with respect. On the other hand, I asked, why should who I am, matter? Because I knew what the pain of sickle cell disease was like, our pain management program was second to none at Baylor. After this experience I knew I had my job cut out for me should I be offered the directors position.

I was able to schedule a later flight home that day. Although not feeling my best, I thought I could handle the flight. I continued my pain medicine by mouth and drank lots of water. Thankfully, the flight home was a direct flight and was uneventful. I arrived home safely and after a day or so I was back to normal only to wait on the decision from the dean.

The two weeks passed slowly but finally the letter came. I was officially offered the position of Division Director of Emergency Medicine at Beth Israel Hospital, Harvard University. I was so happy. This was the beginning of the rest of my life. I was to begin in three short months. I could hardly wait.

I spent much of the next three months preparing for the move to Boston. It is amazing how much stuff one can accumulate over the years. To my surprise I found myself a little sad about leaving Houston, leaving Baylor, and leaving many old friends. None-the-less, that day came and the movers packed my office and apartment. I was on my way to start the next phase of my career at a place where my career was once in its adolescence. As I drove across country, my mind was filled with my own experience in the emergency room. I asked myself, how do I implement a positive change in my staff's attitudes on effective pain management without alienating them? The prevailing negative attitudes about the treatment of sickle cell pain crisis are universal. This was going to be a challenge.

That magical day came when I started as the new Division Director of Emergency Medicine at Beth Israel. When I entered the emergency room that morning, there was a stillness across the room. I spoke to each of the staff by name as I passed them. They looked in bewilderment. I immediately spotted the resident that had treated me for my pain crisis. I could tell he did not recognize me. As I entered my office, I could here them mumbling, most with smiles on their faces. For some that was their first time seeing me. I think it was still hard for some of them to believe that this five foot- two-inch, bronze skin woman with an afro, was their new director. I wasn't in the office for more than an hour before a three-car accident resulted in seven severely injured people entering the emergency room. I entered the patient area and began triaging patients based on severity as the staff broke up into teams. As life would have it, the resident who cared for me was on my team. The patient we had was being bagged on 100% oxygen, had a flailed chest and was hypotensive. The patient's airway was intubated through the nose, two large bore IV's were established and the chest was stabilized. He subsequently developed a malignant heart rhythm that required the patient to be shocked to establish a normal rhythm. After being cardioverted, the patient was stable but critical and we were able to transfer the patient to the intensive care unit for further evaluation and management. Shortly thereafter, things quieted down in the emergency room. The resident looked at me with a big smile on his face and said, "you really are as good as they said you are." I smiled and went on. Before I left I told him to come by my office the following day so we could talk. He said I'll be there and went on to see some of the other patients.

The following day the resident came by as we had discussed. As he entered we began by discussing his future plans. He, like I once was, was ranked as one of the top residents. He someday wanted to head his own division. Without warning he said I couldn't help but think we have met before. I said, "We have. Do you remember treating a lady about three and a half months ago for a sickle cell pain crisis that looked like me?" He stared closely and mumbled, "yes, but no, you're that lady?" "Yes."

"You've got sickle cell disease?" "Yes." "Who would have thought?" "Let this be a lesson, always treat your patients the way you want to be treated. You never know they may be your doctors some day."

Written by
Johnson Haynes, Jr., MD
August 8, 1999

*Note: The characters in this narrative, "Who Would Have Thought?" are fictional and was written to address attitudes reflected by sickle cell disease clients experienced globally in healthcare settings.*