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# Sickle Cell Today
## USA Comprehensive Sickle Cell Center

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# Remarkable Decline in Pneumococcal Infection in Children with Sickle Cell Disease at Children's & Women's Hospital

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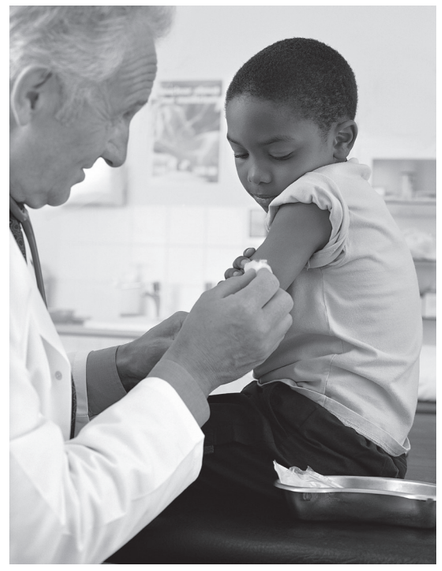

Multiple factors have played a role in reducing the rate of pneumococcal infections in children with sickle cell disease. A large clinical trial conducted in 1986, concluded that penicillin prophylaxis for children younger than 6 years, with sickle cell disease, was protective. In addition, Pneumovax (a vaccine against 23 varieties of pneumococcal infections) which is administered at 2 and 5 years of age, plays a key role in reducing the infection rate. This vaccine was previously shown not to be effective in children younger than 2 years. In 2000, a new vaccine called PCV7 was introduced and was administered to all children at 2, 4, 6 and 12-15 months of ages. It is called PCV7 because it protects against 7 different varieties of pneumococcal bacteria. More recently, in 2010, the 13-valent pneumococcal polysaccharide conjugate vaccine (PCV13) was introduced to replace the PCV7 for all children.

***see "Decline" on page 4***

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

*FROM THE DIRECTORS DESK:*

# *ONE BAD APPLE DOESN'T SPOIL THE WHOLE BARREL*

It had been an exceptionally warm fall for the month of November in Alabama. Everyone was still running around in shorts and short sleeves. I was just about to finish my shift in the emergency room. All of the doctors were commenting on what an unusually quiet month it had been except for the routine chest pains, chronic "lungers," diabetics, and the "frequent flyers" with sickle cell disease that seemed to live in the emergency room most days of the month. It was a standing joke amongst the doctors how two patients with sickle cell disease, Margaret and Bobby, were in the emergency room for their daily shots of demerol and how they were just addicts. Every time I heard this it would make me so angry. I would often wish they could just walk a mile in Margaret's and Bobby's shoes. Mary and I were the only ones who were willing to treat them appropriately and respectfully when they came to the emergency room. Some of my colleagues would even try giving them sterile water for pain. All I knew was as doctors, we had no means to objectively measure pain and it always boiled down to believing the patient. It often seemed so unfair. Maybe they were not having a pain crisis and maybe they were just asking for help. All I knew was if it were me hurting, I would be very angry if I had to beg for the treatment I deserved if I had to convince someone I was in pain.

With only two more days to go on the emergency room rotation there was a sudden change in temperature. When I went to bed that night it was 75 degrees F. The following morning the temperature had dropped to 28 degrees F. I knew when I got to work the emergency room was going to be full. It always was following a sudden change in the weather. When I arrived to work it was as expected: all the exam rooms were filled with chronic "lungers" who could not breathe, and at least six patients writhing in pain with sickle cell disease. Amongst the sickle cell patients were Margaret and Bobby. The other four patients were not any with whom I was familiar. When the night shift doctor checked out to me he said, "this place is full today with more of those sicklers. They really aren't hurting. They just want more demerol. That sickler's arms over there are full of track marks from shooting up crack-cocaine. There are too many addicts in here for me. Good luck." I proceeded to evaluate the patients. Other than Margaret and Bobby, none of the other patients with sickle cell disease had been admitted to the hospital or emergency room for over a year and one of them had experienced only two crises in his life. Despite all of this, only Mary and I were willing to treat the sickle cell patients. We ended up admitting two of them for chest syndrome with pain crisis and one young lady for a kidney infection and pain. After several hours, the others were discharged home and given prescriptions for pain medications. Just as the resident had suggested, the patient that used crack-cocaine ended up altering his prescription before having it filled so he could get three times the prescribed number of pills. As life would have it, the local pharmacist called to verify the prescription and the true addict was caught. The talk in the emergency room the next day was all about this. No one spoke of the two patients who had chest syndrome, one of whom had to be emergently placed on the ventilator, or the patient who had the kidney infection, or the patients who were discharged home.

One bad apple doesn't spoil the whole barrel. None the less, this is not how many physicians see it. They see all patients with sickle cell disease as addicts, as liars about their pain. How do I get my colleagues to see individuals with sickle cell disease individually? Why is it that we as physicians are so quick to judge others as addicts when addiction amongst us is more prevalent than that seen amongst those, whom we judge? What if individuals with sickle cell disease judged us using the same bias?

Written by
Johnson Haynes, Jr., MD

**Note: The characters in this narrative, "One Bad Apple Doesn't Spoil the Whole Barrel" are fictional. The narrative was written to address attitudes commonly reflected in healthcare settings.**



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## MARK YOUR CALENDAR

# THE 2014 USA ANNUAL SICKLE CELL CONFERENCE:
## Practical Issues XIII-The Many Faces of Pain

The USA Comprehensive Sickle Cell Center is in the final stages of planning its 13th dynamic and informative Annual Regional Sickle Cell Conference. The focus this year will be on **"Pain in Sickle Cell Disease and Pain Management."** The conference is scheduled for Saturday, May 3, 2014 from 8:00am – 3:45pm in the School of Allied Health auditorium on the main campus of the University of South Alabama. National and local experts will be presenting the most current and practical issues experienced by healthcare providers caring for those affected by sickle cell disease and pain management. The target audience is physicians, physician-assistants, nurse practitioners, nurses, and allied health professionals. This conference is supported by the Dr. Cecil L. Parker Jr. Sickle Cell Disease Lectureship Endowment. The purpose of the Endowment is to provide support for the Annual Regional Sickle Cell Conference and educational needs of the clients and health care providers of the Gulf Coast community.

Register early to enter for a chance to win complimentary admission to the 2015 Annual Regional Sickle Cell Conference. Early bird registration deadline is April 11, 2014. For additional conference information visit http://www.usahealthsystem.com/newspecial-events or call (251) 470-5893.

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***References continued from page 3***


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# *Sickle Cell Disease and Hydroxyurea: Yesterday, Today, and Tomorrow*

*Submitted by*
*Ardie Pack-Mabien, CRNP*

Mandatory newborn screening in the United States has resulted in early identification and medical access by those newborns affected with sickle cell disease (SCD). The addition of penicillin prophylaxis and pneumococcal immunizations as preventive measures, technological advancements in the development of diagnostics and screening tools and innovative medical management of the pediatric population have resulted in decreased morbidity and mortality for individuals living with SCD. The improvement in survivorship from childhood to adulthood has been estimated at 94% for individuals with HbSS and HbSβ$^0$ Thalassemia and 98% for individuals with HbSC and HbSβ$^+$ Thalassemia (Quinn, et. al, 2010).

What role has hydroxyurea played in the medical management of individuals with SCD? The utilization of hydroxyurea as a potentially useful drug therapy in the management of individuals with SCD was first reported in 1984. Additional multi-center studies sponsored by the National Institutes of Health were conducted and found that hydroxyurea reduced the frequency and severity of sickle cell related pain crisis, number of acute chest syndrome episodes, frequency and number of red blood cell transfusions, and decreased end organ damage (Platt et al., 1984, Charache et al., 1992, Charache et al., 1994). Hydroxyurea was approved by the U.S. Food and Drug Administration for the treatment of adults with sickle cell anemia (HbSS) and HbSβ null Thalassemia in 1998 but has not been FDA approved for children with SCD. Since its approval, ≈ 40% of the adult population and 10% of the pediatric population at the University of South Alabama Comprehensive Sickle Cell Center are currently taking hydroxyurea for the management of their disease. What we have learned over the last decades is that hydroxyurea reduces morbidity and mortality for children and adults with sickle cell anemia and it is cost effective. It has been shown to reduce emergency room visits and hospital admissions, and the need for red blood cell transfusions. Hydroxyurea is well tolerated without significant short term toxicities or long term safety concerns. Its efficacy in SCD has been demonstrated in a large body of evidence-based research, and based on disease severity, should be considered as a possible disease modifying therapy in all patients with SCD (McGann & Ware, 2011). Hankins et. al. 2005, have previously reported that infants treated with hydroxyurea therapy have improved splenic function and growth rates, tolerated prolonged hydroxyurea therapy with hematological benefits, fewer acute chest syndrome episodes, and possibly preserved organ function. Safety remains a concern for health care providers, patients, and parents of children, thus individuals taking hydroxyurea therapy should be followed by their hematologist or sickle cell specialist in conjunction with their primary care provider for routine clinical evaluation and laboratory tests to monitor for potential side effects on a regular scheduled basis.

While evidence has mounted over the last decades supporting the long-term safety and efficacy of hydroxyurea therapy as a standard of care for patients with SCD (McGann & Ware, 2011), the question remains, at what point should hydroxyurea be considered the standard of care for all individuals with SCD (HbSS, HbSβ-thalassemia, and HbSC disease).

**References continued on page 6**

# *PLAYING IT SAFE*

The Center for Disease Control and the Advisory Committee on Immunization Practices are now recommending that individuals 19 years or older with sickle cell disease receive the Prevnar 13 vaccine. Prevnar 13 was implemented in the pediatric population in 2010. While Pneumovax has been found to be effective against potentially life threatening infections due to the pneumococcal bacteria, Prevnar 13 vaccine has the ability to provide additional protection. Please see the sickle cell nurse or contact your healthcare provider for information on the vaccine.

*Submitted by: Brittany Brown, BSN, RN*

***"Decline" continued from page 1***

Between January 2006 and June 2012, a study led by Dr. Abdul Hafeez Siddiqui was conducted at the University of South Alabama Children's and Women's Hospital which reviewed 456 hospitalizations in 133 children with sickle cell disease admitted with fever. Of the 456 blood samples drawn, 19(4%) samples grew bacteria and all of these patients were successfully treated with appropriate antibiotics. Only 2 (0.4%) cases of blood stream infection from Pneumococcus were seen. Interestingly, both these infections occurred before 2010 which marks the introduction of PCV13 vaccine. In other words, there has not been a single case of pneumococcal infection since the start of PCV13. In the past, over 10% of children became infected with the Pneumococcal bacteria and more than a third of them died before their 5th birthday. This study marks a dramatic drop in blood stream infection due to Pneumococcus in children with sickle cell disease in our institution when compared to previous reports.

In this study it was also discovered that the blood culture in almost all of the patients with a blood stream infection turned positive within the first 24 hours of collection. Based on this finding, selected patients are now discharged home after 24 hours of hospitalization with close follow up, instead of 48 hours. Among other causes of fever, a fourth of the patients presenting with fever either had pneumonia or acute chest syndrome. About 13% of these patients tested positive for viruses and Influenza (flu virus) was the most common culprit.

The results of this study were presented at the annual meeting of the American Society of Hematology (ASH) in Atlanta, GA held in December, 2012 and the manuscript was published in the Journal of Pediatric Hematology and Oncology, 30:432-436, 2013.

Doctors at the University of South Alabama Comprehensive Sickle Cell Center Pediatric Clinic, held at Children's and Women's Hospital, are vigilant in assuring strict adherence to the recommended immunization schedule and penicillin prophylaxis. In addition, the parents are educated to watch their children closely for fevers. If the body temperature rises above 101F, parents are advised to bring their child immediately to the emergency room. In the emergency room blood cultures are drawn and patients are admitted to the hospital for antibiotics through veins for at least 48 hours. Further management is decided based on the results of blood cultures and clinical assessment. With this approach and the parents' cooperation, a remarkable decline in blood stream infection due to the Pneumococcus bacteria in children with sickle cell disease has been realized.

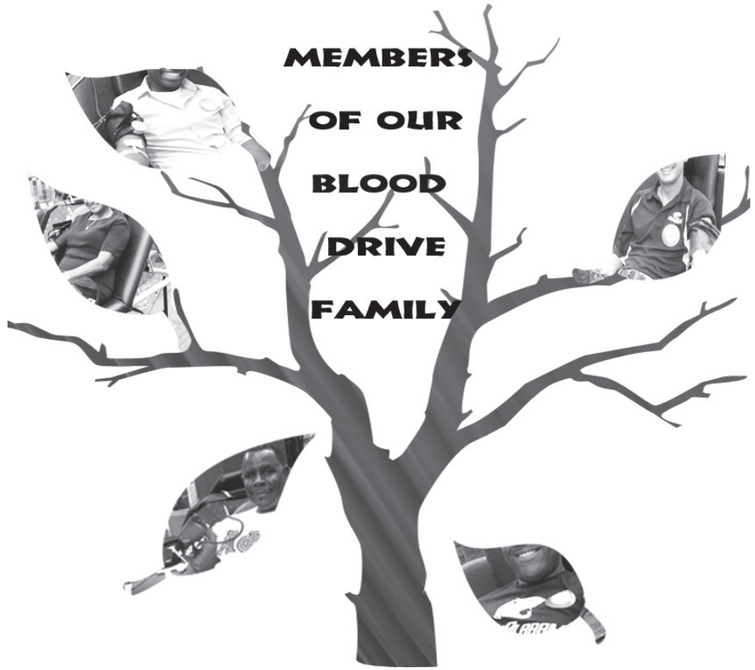


# A Record Breaking Year for the 2013 Annual Blood Drive: Mobile Showed Up and Showed Out!

*Submitted by: Ardie Pack-Mabien, CRNP*

The USA Comprehensive Sickle Cell Center, Alpha Phi Alpha Fraternity, Inc., Franklin Primary Health Center, and the Sickle Cell Disease Association of America, Mobile Chapter 2013 Blood Drive was a **HUGE** success. This partnership began 8 years ago and is conducted annually during the month of September in recognition of National Sickle Cell Awareness Month. The 2013 blood drive was held on Saturday, September 21, 2013 at the Franklin Memorial Complex Mall located at 1303 Martin Luther King Avenue, Mobile, Alabama.

Over the years, many lives in the community have been affected by this local blood drive and each year the blood drive has grown in participant number and success. The blood drive for 2013 had the largest turn-out and shortest wait time compared to previous years. There was an overwhelming amount of support from the Mobile community at the 2013 blood drive. **The blood drive supporters included local, community organizations, medical and physician assistant students from the University of South Alabama, and the Alpha Elites**. Community volunteer efforts included registering donors, serving refreshments, prize distribution, and blood donation. A record breaking 72 individuals presented as possible blood donors. The goal of this blood drive was to collect 45 units. This goal was exceeded and 59 units of blood were collected. Three donors gave what was equivalent to two units using the ALYX procedure. ***Each unit of blood obtained was separated into red cells and plasma possibly touching the lives of 177 individuals***.




The community and local organizations competitive spirit ran high at the 2013 blood drive, as the Classic Corvette Club dethroned the Pacesetters Motorcycle Club with the largest number of blood donors. This was no easy task in that Pacesetters Motorcycle Club has held this honor for the last 3 consecutive years. "Way to Go Classic Corvette Club". The challenge extended today is, who will be the winner at the 2014 blood drive?

The sponsors would like to thank the community, local organizations and volunteers who participated in the 2013 blood drive. ***Thank you for your cooperative spirit, dedication, and continued support. Thank you 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 2014 blood drive.

The 2014 Blood Drive is tentatively scheduled for Saturday, September 20, 2014 at Franklin Primary Health Center located at 1303 MLK Drive, Mobile, Alabama.