

Sickle Cell Today

USA Comprehensive Sickle Cell Center

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Bridging The Gap

Through The Pediatric To Adult Care Transition Program



PNC Bank Grant Supports PACT Program. Left to right Cedric Hatcher, PNC Regional President; Johnson Haynes, Jr., M.D.; Danisha Maye, PNC Community Consultant

T'Shemika Perryman, RN – PACT Coordinator

Since 2012, the Pediatric to Adult Care Transition Program (PACT) has bridged the gap between pediatric and adult healthcare delivery for clients with sickle cell disease. Now, a longtime goal is close: PACT will expand to include the Learning and Resource Development Center (LRDC), a facility that will be equipped to assist clients in acquiring the skills they need to live full and productive lives.

PACT has already helped young adult patients to make the transition to an often-frightening adult system. “The program helped me step-by step to plan out my next steps to adult care and make my adult transition easier,” said 19 year-old Brianna Kennedy, a client with sickle cell disease.

In the last four years, sixty-six pediatric clients between the ages of thirteen and nineteen, all with sickle cell disease, have enrolled in PACT; 22 have successfully transitioned to adult

see “*Bridging*” on page 5

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

IS YOUR CHILD ON HYDROXYUREA ?

Hamayun Imran, MD – Medical Director, Division of Pediatric Hematology/Oncology

For many hematologists, sickle cell anemia (SCA) is correctly defined as a chronic and progressively debilitating medical condition with deceptive but inevitable organ damage. Pediatric patients may appear to be healthier than they really are to their families and healthcare providers if they are not experiencing frequent pain episodes or recurrent hospitalizations. End organ damage not commonly recognized in children with SCA are delayed growth and development, neurocognitive delay, poor school performance, brain vessel damage and splenic and kidney dysfunction. Therefore, early preventative treatment in children who have not yet developed serious or irreversible organ damage is of paramount importance.

Hydroxyurea (HU) is the only medical therapy other than red blood cells transfusions proven to prevent known complications of SCA. In 1988, the Food and Drug Administration approved the use of HU in adults with SCA whose course was complicated by frequent pain crises and recurring acute chest syndrome. The National Institutes of Health (NIH) Office of Medical Applications of Research (OMAR) and the Agency for Healthcare Research and Quality (AHRQ), after a comprehensive review of the evidence, have concluded that HU is also efficacious in children with SCA. As seen in adults, HU increases the hemoglobin level and the percentage of fetal hemoglobin along with a concurrent reduction in total white blood cell, platelet and reticulocyte counts. These effects on blood cells have been clinically found to reduce pain episodes, acute chest syndrome, hospitalizations, need for blood transfusions and likely prevention of organ damage.

Evidence Supporting HU in Children with Sickle Cell Anemia. In children with SCA, the Hydroxyurea Safety and Organ Toxicity (HUSOFT)

trial reported that infants tolerate HU without short-term toxicity and have substantial laboratory and clinical efficacy. The HUG-KIDS study reported normal growth and development and sustained laboratory benefits for infants and school age children. The BABY HUG trial did not show clear benefits for organ protection in infants but did demonstrate a significant reduction in pain crises, acute chest syndrome, hospitalizations, and transfusion therapy. The National Heart Lung and Blood Institute sponsored clinical trial, Stroke With Transfusions Changing to Hydroxyurea (SWITCH), demonstrated a lack of effectiveness of HU in children having had a stroke and that red blood cell transfusions and chelation remain a better way to manage children with SCA, stroke, and iron overload. In contrast, the Transcranial Doppler with Transfusions Changing to Hydroxyurea (TWITCH) clinical trial clearly showed that HU is as effective as red blood cell transfusions in children with SCA who have not had a stroke but are at high risk for having a stroke as demonstrated by an abnormal transcranial doppler velocity.

HU Therapy and Potential Adverse Effects. It is a well-tolerated medication that is taken by mouth and is available in liquid and tablet. It may cause an occasional upset stomach that can be overcome by changing daily dosing to evening administration. Darkening of the nails and skin may occur but hair loss is rare. Transient and reversible suppression of blood counts can occur. Thus, complete blood counts should be monitored closely, ideally at monthly intervals, particularly when starting HU therapy and escalating the dose. While leukemia has been reported to be a rare (<5%) occurrence in patients treated with HU, a small teenage cohort from the HUSOFT study who were followed over a 14 year period of treatment showed no evidence of leukemia

while taking HU and had good health with normal growth and development. Furthermore, the NIH consensus statement regarding HU treatment states that “the available evidence does not support the association of HU treatment with the development of leukemia in adults or children.” Lastly, because of concerns about the potential of HU to cause birth defects, the drug is generally not prescribed to pregnant females.

In summary, HU is an important therapeutic option for children and adolescents with SCA and recent evidence support long-term benefits with prevention of chronic organ damage. However, due to inadequate provider and family knowledge about HU, patients are either not offered HU or treatment is declined because of unfounded fears. Social issues, healthcare structure and limited support for HU by nonprofessional organizations also contribute to its underuse. Therefore, despite extensive published evidence, HU still is not being widely prescribed. We must overcome these barriers otherwise this potentially disease modifying therapy will continue to be underutilized. As pediatric sickle cell providers, we are obligated to provide our children with SCA and their parents with the most up-to-date information on the best available therapy for their potentially incapacitating and progressive illness. By doing this, our patients and their families can make informed decisions. While hematopoietic stem cell transplantation can be curative, its use and availability are limited due to a variety of reasons. Therefore, at this time, HU appears to be the best available treatment option. In short, it’s time we all consider HU therapy as the standard of care for patients with SCA as young as 6 months of age. At the next visit, please ask your child’s sickle cell provider, why isn’t your child on HU!

2015-2016 Flu Season Will Be Here Soon: How Well Does The Flu Vaccine Work?

Ardie Pack-Mabien, CRNP

Researchers and scientists of the World Health Organization (WHO) meet each year to determine the virus composition of the influenza vaccine for the upcoming influenza season. The influenza vaccine composition is updated each year based on the previous influenza season, how the virus spreads, and which virus made individuals sick. The U.S. Advisory Committee on Immunization Practices (ACIP) votes to make recommendations for 2015–2016 based on the findings of the WHO. The Centers for Disease Control and Prevention (CDC) must approve the ACIP recommendations before being released to health care providers, public health departments, and the public. Although it is not possible to predict exactly what type of influenza season 2015-2016 will bring to the general public, the CDC and researchers conclude that when the composition of the influenza vaccine match well with the circulating influenza viruses, the influenza vaccine is beneficial for at-risk individuals and the general public. The 2015–2016 influenza vaccine virus composition for the Northern Hemisphere includes: a) H1N1, b) H3N2, and c) B/Phuket/3073/2013-like virus.

The influenza season typically occurs between October and May and usually peaks in the United States between December and February. A yearly influenza vaccination is recommended by the CDC for all persons with sickle cell disease ages 6 months and older who do not have contraindications. The influenza vaccine is particularly important for individuals who are at risk for severe complications associated with the influenza virus. Adults and children with sickle cell disease who have chronic lung disease (including asthma), heart disease, kidney, and liver conditions, along with diabetes, stroke, or loss of spleen function should receive their annual influenza vaccine.

It is recommended that health care providers begin offering the influenza vaccine soon after it becomes available and if possible by October to ensure protection during the influenza season. Vaccines are generally available through the month of May or as long as the influenza virus is circulating throughout the community. Children ages 6 months through 8 years who are receiving the influenza vaccination for the first time should receive two doses of the vaccine at least four weeks apart. The influenza vaccine is offered by many health care providers, health departments, clinics, pharmacies, college health services, and employers. See your health



care provider sooner rather than later; the supply of the influenza vaccine may be limited so you may miss out on the possible benefits of the influenza vaccine. Individuals with the flu often miss days from work or school, pay costly copays for medical visits and medications, and may spread the virus to family members and the general public.

Help prevent the spread of the flu by following the recommendations listed below:

- Proper handwashing,
- Turn your head and cough or sneeze into the sleeve of your elbow or napkin,
- Stay at home if you are sick with the flu,
- See your health care provider for your influenza vaccine, and
- Contact your health care provider for flu-like symptoms:
 - Cough
 - Sore Throat
 - Runny Nose, Stuffiness or Congestion
 - Fever
 - Fatigue
 - Headache or Body Aches
 - Diarrhea and vomiting although more common in children

For more information: <http://www.cdc.gov/flu74>

USA SICKLE CELL CENTER: LEGACY, LEADERSHIP AND LEARNING

Johnson Haynes, Jr., MD –Director, Comprehensive Sickle Cell Center

This year's sickle cell conference, *Sickle Cell Disease Practical Issues XIV: The Good, The Bad, The Misunderstood*, was exceptional and well-attended. The annual conference, sponsored by the USA Sickle Cell Center, dispenses up-to-date information and advice about best practices to healthcare providers and patients alike.

The keynote speaker at the May gathering, Ms. Wanda Borders, L.B.S.W., talked about the significance of the social worker's role as an educator, counselor, case manager, mediator, advocate and liaison in optimizing patient care and promoting hope. She is the Associate Director of the Central Alabama Chapter of the Sickle Cell Disease Association of America, based in Birmingham, Alabama.

Felicia Wilson, M.D., Professor, Department of Pediatrics, Division of Hematology/ Oncology, University of South Alabama, addressed the general topic of sickle cell trait and raised the question of whether sickle cell trait is benign when conditions such as splenic infarction, renal medullary carcinoma and sudden death are clearly associated with it.

Sudden death in athletes with sickle cell trait was explored by Lynn Batten, M.D., Associate Professor, Department of Pediatrics, Division of Cardiology, University of South Alabama. Dr. Batten reviewed the potential role of "exertional sickling" and cardiac conditions associated with sudden death in athletes. She also reviewed the current National Collegiate Athletic Association requirement that all athletes be tested for sickle cell trait, despite objections from the American Society of Hematology.

Carolyn O'Bryan Miller, L.C.S.W., PIP, Associate Director, Project Development and Quality Improvement, Managed Care Division, Alabama Medicaid, addressed coming changes in the way that Medicaid services will be provided. With this shift in service delivery to Regional Care Organizations, a greater emphasis on prevention, quality initiatives and treatment is expected to result in an improvement in the quality of care.

Pulmonary hypertension (PH) is an increasingly recognized complication of sickle cell disease and has been reported to cause significant morbidity and mortality. Karen Fagan, M.D., Professor of Medicine and Director of Pulmonary and Critical Care Medicine, University of South Alabama, provided an update on pulmonary hypertension in sickle cell disease. Dr. Fagan emphasized the lack of specificity of the tricuspid regurgitant jet velocity of > 2.5 m/s obtained by Doppler echocardiography in diagnosing PH; she also emphasized that right heart catheterization remains the gold standard in diagnosis.

The most common reason for hospitalization in sickle

cell disease is pain crisis, which often requires the judicious use of opioid analgesics for effective management. Jack A. DiPalma, M.D., Professor of Medicine and Director of Gastroenterology and the Digestive Health Center, University of South Alabama, spoke on opioid-induced constipation. Dr. DiPalma emphasized that constipation is not a trivial problem and that many of our patients suffer in silence.

Errol Crook, M.D., Professor and Chair, Department of Internal Medicine, University of South Alabama, provided an update on chronic kidney disorders in sickle cell disease. He emphasized that "renal disease in sickle cell is common, but it may not be commonly recognized." He offered a comprehensive review, addressing disorders affecting the kidney in sickle cell disease, as well as early diagnosis and treatment strategies.

Attendees of the conference, held on May 2, 2015 at the University of South Alabama Medical Center, received up to 7 *AMA PRA Category 1 Credits™*.

Ms. Wanda Borders, the keynote speaker, was the seventh recipient of the Dr. Cecil L. Parker, Jr., Sickle Cell Disease Distinguished Lectureship Endowment Award. **Support of the Dr. Cecil L. Parker, Jr., Sickle Cell Disease Distinguished Lectureship Endowment will assure the continued provision of high-quality education for patients and healthcare providers in Mobile and surrounding areas.**

Ms. Marilyn Chancellor, Administration Assistant, received an award for eight years of exemplary service to the USA Sickle Cell Center. The USA Sickle Cell Center has provided over three decades of educational leadership for Mobile and surrounding counties. The next conference is tentatively scheduled for April 30, 2016. Please mark your calendar. Hope to see you there.



2015

Wanda Borders, LBSW
Keynote Speaker
Johnson Haynes, Jr., MD

Johnson Haynes, Jr., MD
Marilyn Chancellor
Cecil L. Parker, Jr., MD

Scholarly Activity

GRADUATES

The University of South Alabama Comprehensive Sickle Cell Center encourages all of the graduates to continue your education and/or training and want to congratulate you on a job well done. The following have successfully completed high school, technical school, college or a career development program.

- * Tracy Logan
- * Tarane Robinson
- * Jomyron Brown
- * Jonathan Suero

Articles Published

1. Pack-Mabien, A., Brown, B., Herbert, D. E. and Haynes, J. (2015), Iron overload in adults with sickle cell disease who have received intermittent red blood cell transfusions. *American Assoc Nurse Prac.* doi: 10.1002/2327-6924.12221.
2. Quarmyne M-O, Mpollo MS, Vanderah L, Canter C, Rayes O, Gonsalves C, Haynes J, Clavijio C, Uwe C, Kalinyak K, Joiner CH, Vinks AA and Malik P. A Phase I Trial of Zileuton in Sickle Cell Disease. *Blood* 2013.Nov; 122(21):993. Available from <http://bloodjournal.hematologylibrary.org/content/122/21/993/>
3. Wood JC, Pressel S, Rogers ZR, Odame I, Kwiatkowski JL, Lee MT, Owen WC, Cohen AR, St. Pierre T, Heeney MM, Schultz WH, Davis BR, Ware RE and for the TWITCH Investigators. Liver Iron Concentration Measurements by MRI in Chronically Transfused Children with Sickle Cell Anemia: Baseline Results from the TWITCH Trial *Amer J Hematol* <http://onlinelibrary.wiley.com/doi/10.1002/ajh.24089/abstract>



2015 Annual Blood Drive Sets Bar Higher- Goal of 60 Units!!

Saturday, September 19, 2015 will mark the tenth year that Alpha Phi Alpha Fraternity, the USA Comprehensive Sickle Cell Center, Franklin Primary Health Center and the Sickle Cell Disease Association of America, Mobile Chapter, have partnered to sponsor one of the most successful blood drives undertaken in the Mobile area.

In the first few years, the coalition of partners set a goal of 25 units of blood. In 2010, the group increased the goal to 50 units. From 2010-2014, a total of 255 units of blood were collected — an average of 51 units per year.

The continued success of this blood drive reflects the power, commitment and level of accomplishment seen when communities work together. The blood drive is held each year in September to commemorate *National Sickle Cell Awareness Month*.

On September 19, the blood drive will be held at Franklin Primary Health Center, located at 1303 Martin Luther King Drive, Mobile, Alabama. Please come and participate by giving the “Gift of Life” through blood donation.



“Bridging” continued from page 1

services.

The LRDC will provide clients with access to laptops, videos, games, and reading materials to further educate them about sickle cell disease. They will also have access to web-based tutorials for ACT/SAT test prep, resume-writing, college applications, and job applications.

The project is led by T’Shemika Perryman, RN, who serves as PACT coordinator, and Ardie Pack-Mabien, CRNP. Drs. Felicia Wilson, Hamayun Imran and Hafeez Siddiqui (Pediatric Hematologist/Oncologist) and Dr. Johnson Haynes, Jr. (Pulmonologist and Adult Sickle Cell provider) make up the physician core of health care providers. Aisha Davis, LBSW, who is with the Sickle Cell Disease Association of America-Mobile Chapter, provides case management services in the pediatric and adult sickle cell clinics.

The PACT program is only possible because of the support of our community partners. Special thanks go to PNC Bank, the Knights of Peter Claver and the Kosmos Club, whose contributions have allowed us to purchase laptop computers for the LRDC. Again, many thanks.

A New Therapeutic Option for Iron Overload in Sickle Cell Disease

Felicia Wilson, MD – Chief of Hematology; Division Director, Pediatric Hematology

Blood transfusions play a major role in the management of acute and chronic complications of sickle cell disease (SCD). It is estimated that over half of children with SCD and nearly 90% of adults with SCD will receive one or more blood transfusions. Although they can be life-saving, multiple transfusions can lead to the accumulation of too much iron in the body. Iron overload (IO), also known as transfusional hemosiderosis, may occur after 10 transfusions. Iron is an essential micronutrient found mostly in red blood cells. It plays a vital role in transporting oxygen throughout the body. Each day through a normal diet, the body absorbs 1 to 2 milligrams (mg) of iron. The body also loses about 1 to 2 mg of iron each day. One unit of blood contains about 200 mg of iron. Therefore, a blood transfusion introduces 100 to 200 times the normal dietary intake of iron into the body over a few hours. There is no physiologic mechanism for the body to get rid of this excess iron.

Once IO occurs, iron begins to accumulate in the heart, liver and endocrine glands including the pituitary, thyroid, pancreas and gonads. If left untreated, serious consequences can develop including irregular heartbeat, heart failure, liver fibrosis and cirrhosis, liver cancer, infertility, growth problems, and diabetes. IO has also been associated with increased pain

crises, organ failure and early death. Unfortunately, there are no warning signs or symptoms for IO. However, it can be diagnosed by a blood test called serum ferritin. Consistent elevations in serum ferritin above 1000 micrograms per liter of blood may indicate iron overload. Because the body cannot get rid of the extra iron, medications referred to as chelation therapy must be used.

The first medication approved for chelation was desferal in 1968. It had to be given as a subcutaneous infusion over 8 to 12 hours after placing a needle under the skin. This procedure had to be repeated 5 to 7 days each week. The inconvenience of a long infusion time, discomfort of repeated needlesticks, and hypersensitive reactions at the infusion site made it problematic for many patients to continue treatment. It would be 37 years before the approval of the oral iron chelator, Exjade in 2005. Although it revolutionized chelation therapy, Exjade had to be dissolved in water, apple juice, or orange juice and taken on an empty stomach once daily. Adverse effects such as nausea, vomiting, diarrhea, and abdominal pain coupled with the consistency of the suspension still made chelation therapy challenging. Health care providers and patients have been eager for alternative options for chelation.

Jadenu, a new formulation of Exjade with the same active ingredient, was approved in March 2015. It is a film-coated tablet that can be taken with or without a light meal once daily. Lactose and sodium lauryl sulfate, thought to cause the

adverse effects of Exjade mentioned above, were left out of the final formulation of Jadenu.

By simplifying treatment administration with potentially fewer side effects, Jadenu is an important new option to help patients meet the goals for successful chelation.

Typically, transfusions are not required just to correct the anemia in SCD. Rather, they are triggered by episodes such as worsening anemia caused by acute splenic sequestration crisis (ASSC) or parvovirus infection, acute chest syndrome (ACS), multi-organ failure, preoperative management and stroke or acute neurologic deficit. These are referred to as episodic transfusions. Some complications require long-term suppression of sickle cells. This is achieved by chronic transfusion protocols that administer transfusions every three to five weeks. Chronic transfusion can reduce the pediatric risk of stroke by 90%. In individuals who have had a stroke, chronic transfusions dramatically reduces the incidence of recurrent stroke from 60% - 90% to less than 10%. Recurrent ACS, chronic renal failure, ASSC and complicated pregnancy all benefit from chronic transfusion. Patients can now take control of their health by being more aware of iron overload, tracking the number of transfusions received, knowing their serum ferritin levels, and adhering to chelation therapy. For more information, visit www.jadenu.com for additional patient and health care provider resources.



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