

Sickle Cell Today

USA Comprehensive Sickle Cell Center

Volume 12 Issue 1

September 2015

Table of Contents

Bridging the Gap Through the Pediatric to Adult Care Transition Program	1
Is Your Child On Hydroxyurea ?	2
2015-2016 Flu Season Will Be Here Soon: How Well Does The Flu Vaccine Work?	3
USA Sickle Cell Center: Legacy, Leadership and Learning	4
Scholarly Activity	5
2015 Annual Blood Drive Sets Bar Higher - Goal of 60 Units!	5
A New Therapeutic Option for Iron Overload in Sickle Cell Disease	6

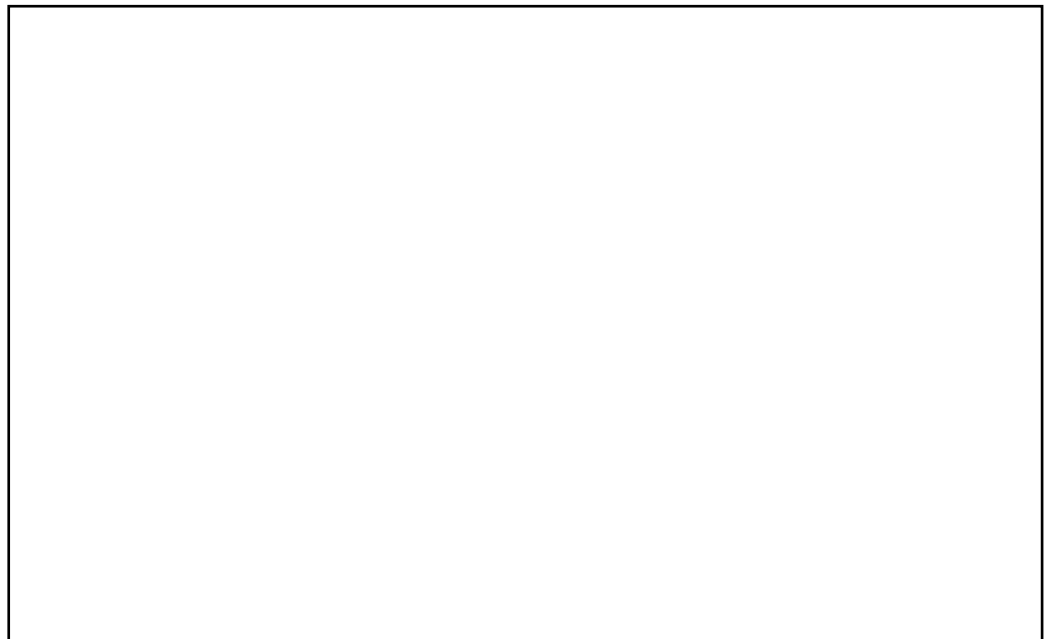
USA Comprehensive Sickle Cell Center

Clinical



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

2015-2016 Flu Season Will Be Here Soon:

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, 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

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

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.

Scholarly Activity

GrADuATeS

Articles Published

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.

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.



Comprehensive Sickle Cell Center

TM

