Sickle Cell Today

USA Comprehensive Sickle Cell Center

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USA Comprehensive Sickle Cell Center
Main Office (251) 470-5893
Fax (251) 470-5895
Clinical Research (251) 471-7703
Pediatric Clinic (251) 405-5147
Adult Clinic (251) 470-5890
Community-based Program (251) 432-0301

TRISICKLE for Sickle Cell

Ardie Pack-Mabien, FNP-BC, PhD

TRISICKLE for Sickle Cell Campaign

As a result of mandatory newborn screening, penicillin prophylaxis, and pneumococcal vaccines, the life expectancy of babies born with sickle cell disease (SCD) in the United States has improved to the 4th and 5th decades of life. While survival with SCD has improved, one-third of adolescents and young adults delay or do not successfully transition to adult care and are lost to medical follow-up for the management of their SCD and its' complications. This improvement in survivorship from childhood to adulthood has resulted in the need to develop programs that foster successful transition and transfer of the adolescent and young adult with SCD to adult care.

To facilitate a more effective transition process and transfer to adult sickle cell disease care, the **Pediatric to Adult Care Transition (PACT) Program**

(continued on pg. 2)

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

TRISICKLE for Sickle Cell (continued)

was implemented at the University of South Alabama Comprehensive Sickle Cell Center on January 6, 2012.

The overall goal of the PACT program is to provide the adolescent and young adult ages 13-19 with a comprehensive patient-centered approach that emphasizes a better understanding of SCD and its' complications, appropriate use and misuse of medications, age-specific insurance benefits particularly with Alabama Medicaid, self-care, self-advocacy, patient-physician collaboration, and interdisciplinary care coordination between pediatric and the adult care providers. The motto of the PACT program is to "em**P**ower and en**A**ble ex**C**ellence **T**ogether" with the participants of this program and their family caregiver(s).

TRISICKLE for Sickle Cell Campaign:
What is it? This effort was birthed by the Sickle Cell Center Development Council which is composed of community leaders, health care providers, clients, and family caregivers. It was developed with the purpose of promoting sickle cell disease awareness and to raise funds in support of programs and services offered by the PACT program and USA Comprehensive Sickle Cell Center.

- In observance of "National Sickle Cell Awareness Month", the *TRISICKLE for Sickle Cell Campaign* had its' kick-off event at the Annual Sickle Cell Disease Blood drive held on Saturday, September 15, 2018. The campaign started off with great enthusiasm but clearly needs the support of the community.
- Take the pledge to TRISICKLE for Sickle Cell; see how far you can ride, and video yourself riding a tricycle.
- Don't forget to share your video on social media with the tag, #TriSickleForSickleCell and invite a friend to try!
- Pledge $1 per foot and help us raise awareness of and funds for Sickle Cell Disease.
- Donation link: visit facebook.com and search TRISICKLE for Sickle Cell Challenge.

For additional information the TRISICKLE for Sickle Cell Campaign and the PACT program, you can call the University of South Alabama Comprehensive Sickle Cell Center at 251 470-5893 or 251 471-7703.

Pediatric to Adult Care Transition (PACT) Parental Involvement

T'Shemika Perryman, RN, Transition Coordinator

The mission of the PACT program is to improve health and literacy of the adolescent and young adult with sickle cell disease through educational programs and a comprehensive, patient-centered, health care delivery system. This program emphasizes self-management, patient-physician collaboration, and interdisciplinary care coordination directed at increasing patient education.

The PACT program focuses on adolescents and young adults between the ages of 13-19 years with sickle cell disease. In this population, the PACT team has identified a lack of transportation availability, interest/motivation, and family support as factors that often impede program participation. Moving forward, the PACT team will promote more parental involvement and input, as it becomes increasingly clearer that parental engagement is essential to the success of the PACT program.

To achieve this goal, on May 30, 2018, the first parent-only meeting was held at the USA Sickle Cell Learning Resource and Development Center. During this meeting, we had several parents to come and discuss what their children's participation in the PACT program meant to them. We also discussed topics ranging from whether or not the PACT program should start earlier than 13 years of age, does any content need to be added or changed, and other avenues that could lead to getting more parental involvement in the PACT program. At the conclusion of this meeting, a decision was made to include a family caregiver on the PACT team. Incorporating parents into the PACT team will hopefully have an overall positive effect on adolescents and young adults involvement in the PACT program.

The Flu season will be here before we know it! Will you be ready?

Ardie Pack-Mabien, FNP-BC, PhD

The dreaded cold and flu season is but a sneeze, cough, and runny nose away. Typically, the flu season begins between the months of October and May and usually peaks in the United States between December and February. The flu season for 2017-2018 peaked during the early part of February with a record breaking hospitalization rate that exceeded prior years.

The influenza (flu) virus is highly contagious and poses a serious risk for individuals living with sickle cell disease. 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, coworkers, and the general public. According to the Influenza Hospital Network Surveillance (FluSurv-Net) Laboratory, there was a cumulative overall rate of 106.6 flu related hospital admission per 100,000 people in the United States for the 2017-2018 influenza season. The highest rates of hospital admissions were among person 65 years of age and older, followed by adults 50-64 years of age, and young children 0-4 years of age (http://gis.cdc.gov/GRASP/Fluview/FluHospRates.html and http://gis.cdc.gov/grasp/fluview/FluHospChars.html). In 2010, flu related illness direct and indirect cost per capita in the United States was an estimated 19.8 million (Peasaha, Azziz-Baumgartnera, Breesea, Melzerb, & Widdowsona, 2010).

To help prevent the spread of this virus, an annual influenza vaccination is recommended for all persons aged ≥6 months unless there are contraindications for the administration of this vaccine. This vaccination is particularly important for individuals who are at an increased risk for severe complications from influenza, or at higher risk for influenza-related outpatient, emergency department, or hospital visits (http://www.cdc.gov/flu/about/season/flu-season-2016-2017.htm). Such individuals include children and adults with sickle cell disease, individuals with diabetes, strokes, loss of splenic function, and chronic conditions involving the lung (including asthma), heart (except isolated high blood pressure), kidney, and liver. See your healthcare provider to discuss the potential risks and benefits of this vaccine.

Health care providers usually begin offering the influenza vaccine soon after it becomes available and continues throughout the month of May or as long as the influenza virus circulates throughout the community. Of note, 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 (http://www.cdc.gov/flu/about/season/flu-season-2016-2017.htm). The vaccine can be obtained from your health care provider, health departments, clinics, urgent care centers, pharmacies, college health services, and employers. See your health care provider sooner rather than later to receive your vaccination as not to miss out on the benefits or possible shortage of this vaccine. Please keep this in mind as the availability and supply of vaccinations may be limited due to growing demands by the general public.

No, the influenza vaccine does not cause an individual to develop the flu. However, there are some short-term and mild side effects of the influenza vaccine (http://www.cdc.gov/flu/about/season/flu-season-2016-2017.htm). That being said, exposure to an individual(s) with the influenza virus prior to receiving the vaccination may increase your risk of developing flu-like symptoms or the flu (http://www.cdc.gov/flu/about/season/flu-season-2016-2017.htm). Potential side effects of the influenza vaccine include: soreness, redness, or swelling at the injection site, low grade fever, and generalized aches.

To help prevent the spread of the flu, the CDC recommends:

- Proper handwashing with soap and water or hand sanitizer
- 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 vaccination
- 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

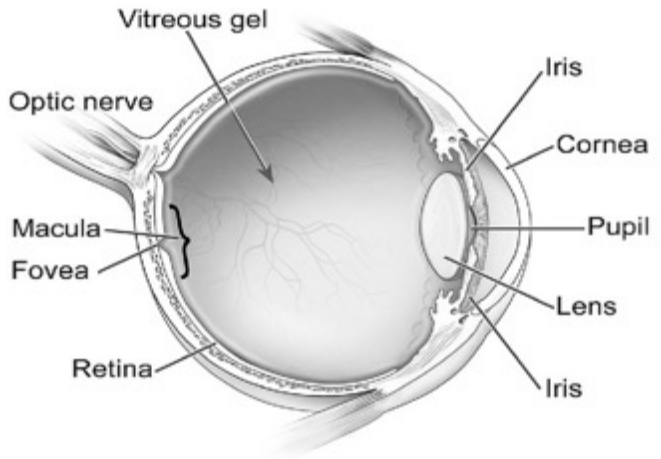
For additional information about the influenza virus, spread, prevention, and vaccine go to the Centers for Disease Control and Prevention website at: http://www.cdc.gov/flu/protect/keyfacts.htm.

Sickle Cell Disease and Eye Health

Jessica L. King, FNP-BC

Individuals with sickle cell disease (SCD) are at an increased risk for eye disease that can lead to permanent blindness if not caught early and treated. All individuals with SCD, especially those with hemoglobin SC, are at risk for injury to the retina. The retina is a layer of cells at the back of the eyeball that are sensitive to light and that trigger nerve impulses that pass through the optic nerve (the nerve that feeds into the back of the eye) to the brain, where sight is formed. Basically, your eyes are like cameras and your retina is the camera's film that gets developed into a picture. In SCD, damage to the retina occurs when rigid, sickled red blood cells get trapped and block blood vessels within the eye. This results in an inadequate blood supply to the eye, retinal injury and subsequent loss of vision, including blindness. When blood flow is inadequate to the eye over a period of time, the eye will make new vessel that grow onto the surface of the retina and/or into the vitreous gel. (See diagram). This can lead to other complications such as bleeding into the back of the eye and retinal detachments. If blood vessels leak and blood cells get stuck within the vitreous gel it can prevent the passage of light through the retina and cause blurred vision. When new vessel growth is detected early at your annual eye examination, laser surgery can be very effective in reducing rates of visual loss and bleeding into the back of the eye.

Since most symptoms of serious eye diseases start out mild, they often go unnoticed until it becomes a medical disaster with the potential to cause permanent blindness. The best preventive strategy for all individuals, not only those affected with sickle cell disease, is having



a dilated exam with an Ophthalmologist once a year. The USA Comprehensive Sickle Cell Center follows the current National Institutes of Health recommendations for individuals with SCD: 1. refer to an ophthalmologist for dilated eye examination to evaluate for retinopathy beginning at age 10; 2. re-screen at 1-2 year intervals for those with a normal retinal examination; and 3. refer individuals with suspected retinopathy to a retinal specialist. Don't wait to test the old proverb, "Seeing is believing". Don't wait to develop something serious before you can accept that it really happens.

Evidence- Based Management of Sickle Cell Disease Expert Panel Report, 2014: Guide to Recommendations. (2014). Retrieved September 15, 2018, from National Institutes of Health Website: http://www.nhlbi.nih.gov/sites/wwEvidw.nhlbi.nih.gov/files/sickle-cell-disease-report.pd

"A Classy Example": Classic Corvette Club of Mobile

The Classic Corvette Club (CCC) epitomizes the ole saying, "you can't lead from behind", both literally and figuratively. Not only do they lead the way in their beautiful fast corvettes, the CCC has been one of the most consistent donors to the University of South Alabama Comprehensive Sickle Cell Center for the last eleven years. In September of every year, National Sickle Cell Awareness Month, this organization has adorned the annual blood drive held at the Franklin Primary Health Center Medical Mall of Mobile, Alabama, with its' beautiful corvettes. And, its' members donate their blood and money in an effort to provide the "gift of life" and to support ongoing educational programs sponsored by the Sickle Cell Center. USA Health and the Sickle Cell Center applaud the CCC of Mobile for being true to its mission, " providing social interactions between members, their families, and the community".

Annual Blood Drive: The Mobile Community Village Gives One Pint at a Time for Sickle Cell

Johnson Haynes, Jr., MD, Director
University of South Alabama
Comprehensive Sickle Cell Center

The annual blood drive sponsored by Alpha Phi Alpha Fraternity, Inc., USA Comprehensive Sickle Cell Center, Sickle Disease Association of America, Mobile Chapter, and Franklin Primary Health Center was held on September 15, 2018 at the Franklin Memorial Complex Mall located at 1303 Martin Luther King Avenue, Mobile Alabama. The blood drive is conducted annually during the month of September, which is National Sickle Cell Awareness Month. 2018 marked the 20th year of the blood drive which is done in conjunction with the American Red Cross.

The success of the drive stems from participation by the Mobile community. Annual supporters of the blood drive are the: Classic Corvette Club, Student National Medical Association/University of South Alabama College of Medicine, Alpha Elites, and donors from across the Mobile community. While the goal of this drive was 50 units of blood, **48** units were collected. **Each unit of blood obtained was separated into red cells and plasma possibly touching the lives of 144 individuals.**

The sponsors express their sincere gratitude to the community, local organizations and volunteers who came out in support of the blood drive. **Thank you for giving the "Gift of Life" through blood donations.** We are looking forward to a bigger and better blood drive in 2019 and hope to see you and your organization present. The 2019 Blood Drive is tentatively scheduled for Saturday, September 14, 2019 at Franklin Primary Health Center Medical Mall located on Martin Luther King Drive, Mobile, Alabama.

USA ANNUAL SICKLE CELL CONFERENCE 2018 (A Legacy of Excellence)

Johnson Haynes, Jr., MD, Director
University of South Alabama
Comprehensive Sickle Cell Center

Sixty-three participants, consisting of physicians, PhD's, a Pharm D, physician assistants, nurse practitioners, registered and licensed practical nurses, social workers, and staff attended the 2018 conference. The 2018 conference, "Sickle Cell Disease Practical Issues XVI: Pain: Pilot or Passenger?," was the 16th 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). The 2018 conference focused on pain, as related to treatment, impact on sleep, and opioid prescribing regulations and requirements.

The Dr. Cecil L Parker, Jr., Sickle Cell Disease Distinguished Lecture was presented by David J. Axelrod, Associate Professor of Medicine and director of the adult sickle cell disease program at Thomas Jefferson University Hospital-Thomas Jefferson University in Philadelphia, Pennsylvania. He lectured on inpatient management of pain crisis in sickle cell disease. Other lecturers featured were local faculty members from University Hospital and Children and Women Hospital. William "Jet" Broughton, Professor of Medicine, Pulmonary/Critical Care Division, addressed sleep medicine with a focus on sickle cell disease and Hamayun Imran, Professor of Pediatric Medicine, Hematology/Oncology Division, discussed the TWiTCH trial which found that high-risk children with sickle cell anemia and abnormal TCD velocities who have received at least 1 year of transfusions and have no MRA-defined severe vasculopathy, hydroxyurea treatment can substitute for chronic transfusions to maintain TCD velocities and help prevent primary stroke. Edwin Rogers, from the Alabama Board of Medical Examiners, provided insight on the new opioid prescribing regulations and requirements. This phase of the conference ended on the topic of patient controlled analgesia in the hospital management of pain which has become the preferred method of opioid administration in many hospitals across the country.

The last phase of the conference was sponsored by the American Society of Addiction Medicine (ASAM) on Risk Evaluation and Mitigation Strategies (REMS). This portion of the conference was free to all attendees and provided 2 AMA PRA Category 1 Credits. This course met the every 2 year, two AMA PRA Category 1 Credits requirements for Alabama Controlled Substances Certificate holders. With full participation in the conference, attendees were eligible to receive 7 AMA PRA Category 1 Credits.

The highlight of the 2018 conference was celebrating the retirement of Representative James Buskey after serving 42 years in the Alabama House of Representatives. Rep. Buskey was presented the Distinguished Service Award from the USA Comprehensive Sickle Cell Center.

Many thanks for the financial support from the USA Health System, Novartis Pharmaceuticals, Global Blood Therapeutics and ASAM. This support has been vital in keeping the cost of meeting registration affordable and has enabled the USA 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 2019.

Pictured left to right are: Drs. Johnson Haynes, Jr, David J. Axelrod, and Cecil L. Parker

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2451 USA Medical Center Drive, MCSB 1530
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