Sickle Cell Today

USA Comprehensive Sickle Cell Center

Volume 7 Issue 1 September is National Sickle Cell Awareness Month September 2010

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USA Comprehensive Sickle Cell Center

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Sickle Cell Trait in Athletes

Sickle cell trait (SCT) is not a disease. SCT is the inheritance of one gene for sickle hemoglobin and one gene for normal hemoglobin. It affects approximately 1 in 8 African Americans and 1 in 1,000-1,400 Hispanics born in the United States. SCT is usually considered a non-life threatening condition but more recent reports question this. In SCT, the kidney's inability to hold on to water normally and the occurrence of blood in the urine are well known and accepted complications. Other conditions seen or associated with SCT are splenic infarction and/or splenic sequestration crisis (usually associated with rapid assent to high altitude), medullary carcinoma of the kidney, and sudden death in military recruits and athletes.

The issue of sudden death in athletes has received much national attention following the death of a 19-year old, Rice University football player in 2006. In the past four decades, 15 football players with SCT have died from "exertional sickling". In the past seven years, death related to SCT has been reported in five college football players in training, two high school athletes (one a 14-year old female basketball player), and two 12-year old boys training for football. As a result of a settlement between Rice University and the football player's family, the National Collegiate Athletic Association Division I Legislative Council decided in April of 2010 that all Division I student-athletes must be tested for SCT, show proof of a prior test or sign a waiver releasing an institution from liability if they decline to be tested. Allowing student-athletes to decline testing for SCT addresses the student-athlete's concern that a positive test for SCT might result in the denial of opportunities to play competitive sports. The new rule will be in effect for the 2010-11 academic year. It should be emphasized that this ruling is not limited to athletes that are African-American, but applies to all athletes.

Athletes with sickle cell trait should not be excluded from participation as precautions can be put into place. The National Athletic Trainers' Association (NATA) encourages institutions to screen incoming student-athletes. The NATA advocates a slow buildup of conditioning activities and frequent rest-and-recovery periods for all student-athletes because this approach can reduce adverse effects caused by SCT and is also a healthier approach overall.

For more information on SCT in the athlete, go to: http://ahsaa.com/ and http://www.ncaa.org/wps/portal/ncaahome?WCM_GLOBAL_CONTEXT=/ncaa/ncaa/academics+and+athletes/personal+welfare/health+and+safety/sicklecelltrait.

Fact of the day: Since the beginning of mandatory newborn screening in Alabama, in 1988, 5-6% of all babies identified with SCT are Caucasian.

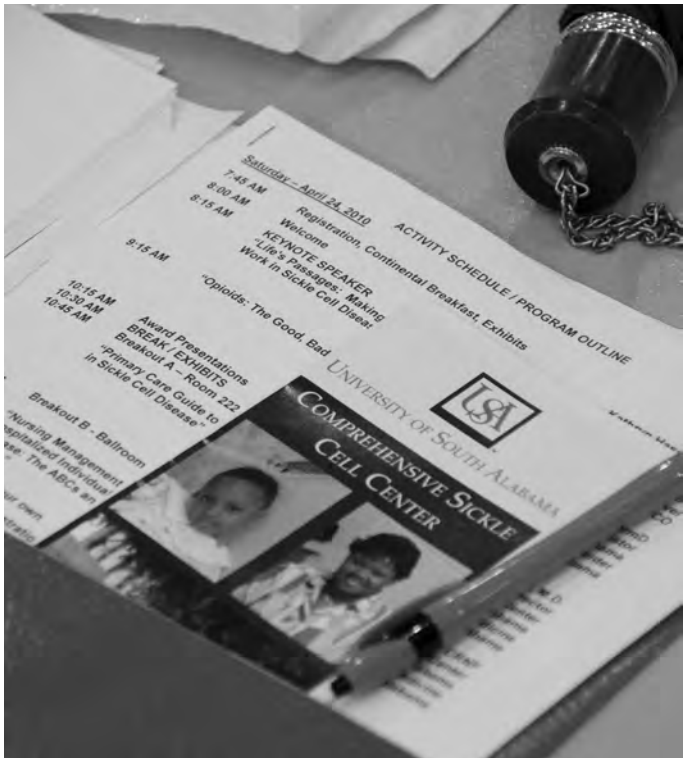
Johnson Haynes, Jr., MD
Professor of Medicine
Director, USA Comp. Sickle Cell Center

Caught in the Net, Despite Weather

"Excellent program, I will be back next year," exclaimed a conference attendee of the 2010 Annual Sickle Cell Regional Conference "Casting The Net". Despite heavy rains, thunder, and the possibility of tornadoes, more than sixty participants consisting of physicians, physician assistants, nurse practitioners, registered and licensed practical nurses, pharmacists, social workers, and staff attended this year's conference. The speakers' evaluations were favorably rated as informative and excellent. The

Dr. Cecil Parker Distinguished Lectureship featured Dr. Kathryn Hassell, a Professor of Medicine and Director of the Sickle Cell Center at the University of Colorado in Denver. Dr. Hassell's lecture addressed "Life Passages: Making Transition Work in Sickle Cell Disease". The conference also featured local speakers from the University of South Alabama: Rachel Weaver, PharmD; Johnson Haynes, Jr., MD; Ardie Pack-Mabien, CRNP; Hamayun Imran, MD, and Benjamin Estrada, MD. Dr. Rachel Weaver, a pharmacist at USA Medical Center discussed "Opioids: The Good, Bad, and Ugly". Dr. Johnson Haynes, Jr., Director of the University of South Alabama Sickle Cell Center addressed guidelines for the primary care physician that could be used in the preventive and acute care of individuals with sickle cell disease. Ms. Ardie Pack-Mabien, CRNP discussed nursing management of the hospitalized individual with sickle cell disease. Dr. Hamayun Imran, Assistant Professor, Department of Pediatric Hematology Oncology discussed splenic sequestration crisis diagnosis and management in sickle cell disease, and Dr. Benjamin Estrada, Professor, Department of Pediatric Infectious Disease provided an update on vaccine preventable diseases in patients with sickle cell disease.

The conference offered the attendees an opportunity to network with other health care professionals from the gulf coast region. Information on the most innovative and up-to-date treatment options in the management of sickle cell disease and its complications were provided by the keynote and conference speakers. Conference attendees thoroughly enjoyed participating in the question and answer sessions.



This conference is held annually in the spring and offers continuing education credits to attendees. Congratulations to Beryl Adkisson, RN winner of the early bird registration drawing for complimentary admission to the 2011 Annual Sickle Cell Regional Conference. For additional conference information and your chance to win complimentary admission for the 2012 conference, call (251) 470-5893.

Ardie Pack-Mabien, CRNP

FROM THE DIRECTOR'S DESK

Good News for Adult Patients Treated with Hydroxyurea!!!!

A study evaluating the long-term efficacy and safety of hydroxyurea in adult patients with sickle cell disease was recently published in the March, 2010 issue of Blood (115:2354-2363). The study was conducted utilizing a single-center trial over a 17 year period. One-hundred-thirty-one patients with sickle cell disease were treated with hydroxyurea and one-hundred-ninety-nine patients received conventional treatment (no hydroxyurea). Patients taking hydroxyurea experienced reductions in the frequency of severe painful crises, transfusion requirements, hospital admissions and the incidence of acute chest syndrome when compared to those receiving conventional treatment. The study further indicated that the 10 year survival probability for patients taking hydroxyurea was 86% compared to only 65% for those patients not taking hydroxyurea. According to the authors, "These results highlight the beneficial effect of hydroxyurea, which appear to modify the natural history of sickle cell disease." The authors further propose to expand hydroxyurea usage to all patients diagnosed with sickle cell disease. The authors have raised an interesting question, should all adult patients with sickle cell disease be treated with hydroxyurea, independent of disease severity? The current recommendations for treatment with hydroxyurea include only adults with sickle cell anemia or sickle β^o -thalassemia who have had 3 or more hospital and/or emergency department admissions in a 12 month period or two or more episodes of acute chest syndromes in a 24 month period. Recent reports have shown that hydroxyurea is underutilized in these populations although clinical benefit has been demonstrated. The data is mounting which supports the premise that early intervention and treatment can result in better long term outcomes and decrease the occurrence of premature deaths in individuals with sickle cell disease. If you are affected with sickle cell disease, see your doctor and ask if hydroxyurea may be beneficial in managing your illness.

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

Hematopoietic Stem-Cell Transplantation in Adults with Sickle Cell Disease: A Chance for Cure

Several studies have reported excellent results with hematopoietic stem cell transplantation (HSCT) in children with stem cells obtained from the bone marrow or from the peripheral blood; however, older patients with severe sickle cell disease (SCD) have not been considered suitable candidates for transplantation. To date, nearly 200 children with severe SCD have been cured with HSCT. In children, complete destruction (myeloablative) of the bone marrow with radiation and chemotherapy are required prior to undergoing HSCT. Such regimens have proven to be too toxic for adults. Adults have years of accumulated organ damage from their disease making the adult less capable of tolerating toxicity from these regimens.

In the December 10, 2009 issue of the New England Journal of Medicine, a group of investigators at the National Institutes of Health reported their results in adult patients with SCD using a "modified blood stem-cell transplant regimen" which did not completely destroy the bone marrow (non-myeloablative). All of these patients were considered treatment failures on hydroxyurea. In contrast to the established method in children, this adult trial sought to reduce toxicity by only partially replacing the bone marrow with donor stem cells. To achieve this goal, the investigators used low dose, whole body radiation and alemtuzumab before transplantation to condition the bone marrow enough to allow donor stem cells to move in and begin producing new, healthy red blood cells. Using this regimen, the patient has to take the immunosuppressive agent, sirolimus, indefinitely after transplantation to suppress the immune system and help prevent rejection of the new stem cells. This relatively low toxicity regimen allowed the patients to become tolerant to the new donor immune cells and to achieve a stable mixture of host (patient) and donor cells called mixed donor–recipient chimerism. Use of this regimen in nine of 10 adults treated resulted in reversal of their SCD. In this study all patients had a HLA- matched sibling donor. Over a 30 month period of follow-up, all are surviving and none of the patients developed acute or chronic graft versus host disease, infertility or worsening of end-organ function. A major limitation of this "modified blood stem-cell transplant regimen" is identification of a HLA- matched sibling donor making this therapy out of reach for all but a small minority of patients. Thus, exploring the use of unrelated stem-cell donors, which is a work in progress, could provide a viable treatment option for an increased number of patients with SCD. In addition, long-term suppression of the immune system carries risk, particularly related to infections and development of the post-transplant lymphoproliferative disorders. Thus, these risks have to be weighed against the risk of complications related to SCD.

For more information related to this study, log on to:
http://soundmedicine.iu.edu/segment/2413/Cure-for-Sickle-Cell-Anemia#

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A Gathering of Family, Friends, and Acquaintances with a Purpose

September 25, 2010
Franklin Primary Health Center
1303 Martin Luther King Drive
10:00 a.m.-2:00 p.m.

The 12th Annual Sickle Cell Center Blood Drive sponsored by Alpha Phi Alpha Fraternity, Inc., USA Comprehensive Sickle Cell Center, Sickle Cell Disease Association of America, Mobile Chapter, and Franklin Primary Health Center will be held Saturday, September 25, 2010 from 10:00 a.m. until 2:00 p.m. at the Franklin Primary Health Center. The Franklin Primary Health Center is located at 1303 Martin Luther King Drive. Forty-three units of blood were collected at last year's blood drive, which positively touched one hundred twenty-five patients, their families, and friends.

Unfortunately, we did not meet our goal of forty-nine units that would have affected a larger number of individuals in need. Your generous support is greatly needed this year in our effort to meet or exceed the goal established for 2010. Please remember, September is "National Sickle Cell Awareness Month". ***Come and participate in the Blood Drive by "Giving The Gift of Life Thru Blood Donation". Please bring a friend.***

Ardie Pack-Mabien, CRNP

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FROM THE SOCIAL WORKER'S CORNER

Hello Everyone! It's "Back to school" time again. Many of our clients classified as seniors will be entering the final phase of their high school education. Some seniors will embrace this milestone with unrelenting hope and determination while others may experience just a tad bit more apprehension. Whatever the case may be, clients interested in pursuing an education beyond high school may be eligible for a full paid scholarship sponsored by The National Sickle Cell Disease Association of America. The "Kermit B. Nash Scholarship" rewards academic excellence and achievement to individuals diagnosed with sickle cell disease. Candidates for this program should be graduating seniors and achieve a G.P.A. of no less than 3.0 unless extreme hardships are demonstrated. Other selection criteria include SAT scores, leadership and community service, extracurricular activities, quality of personal essay, and severity of academic challenges. This is just a brief overview of the various factors involved in the selection process. If you would like more detailed information on the "Kermit B. Nash Scholarship", you may contact me at (251) 432-0301. Until next time, so long from the Social Worker's Corner.

Adrienne Petite, LBSW
SCDAA-MC

Quick Reminder! Flu season is just around the corner.

What should I do to prepare for this flu season? The Center for Disease Control (CDC) recommends a yearly flu vaccine for everyone as the first and most important step in protecting against this serious disease. While there are many different flu viruses, the flu vaccine is designed to protect against the three main flu strains that research indicates will cause the most illness during the flu season. The 2010-2011 flu vaccine will protect against three different flu viruses: an H3N2 virus, an influenza B virus and the H1N1 virus that caused so much illness last season. Getting the flu vaccine soon after it becomes available each year is always a good idea, and the protection you get from vaccination will last throughout the flu season.

What actions can I take to protect myself and my family against the flu this season? The first and most important step in protecting yourself and family against this serious disease is to take your yearly flu vaccine as recommended by the CDC. For information about vaccine supply this season, please visit http://www.cdc.gov/flu/about/qa/vaxsupply.htm.

Contact your primary care provider or local health department for the availability of the flu vaccine and an appointment.

In addition, you can take everyday preventive steps like staying away from sick people and washing your hands to reduce the spread of germs. If you are sick with flu, stay home from work or school to prevent spreading influenza to others..

Who should receive the flu vaccine? Adults and children who have a chronic disorder, requiring medical follow-up or hospitalization due to kidney disease, hemoglobinopathies (sickle cell disease), or conditions that compromise lung function should receive the flu vaccine annually. The flu vaccine promotes immunity to the influenza virus by stimulating specific antibody production.

When should I receive the flu vaccine? The optimal time to receive the flu vaccine is October – November and prior to exposure to the influenza virus. The flu vaccine can be given through the month of December and later as long as the vaccine is available. The flu season may last as long or thru May. There have been shortages of the flu vaccine in the past and it is strongly recommended you do not delay obtaining your flu vaccine.

Reference: www.cdc.gov/flu

Ardie Pack-Mabien, CRNP